HEMOPHILIC ARTHROPATHIES

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Foreword

HEMOPHILIA, A RARE but highly destructive disease, is seen only sporadically, if at all, by orthopaedic surgeons But, for the hemophiliac, the orthopaedic aspects of the illness are of great importance as few sufferers who survive the first years of life are spared the de ction of one or more articulations from hemorrhages into bones

Because surgery was impractical an ative correction of contractures and deformities was recogı in the presence of hemophilia, most orthopaedic surgeo s on bleeders just as general surgeons and dentists wc e their problems. On the other hand, few hemophiliac care because families were ashamed of the strange disease t and wary of publicity Stricken children were kept at hor or wheelchair ambulatory

This situation has been substantially decade by the establishment of The National Hemophi hich attempted to seek out and organize hemophilia suff. he country At the same time, research in hematology est e importance of the use of fresh whole blood to stop bleeding in hemophilia temporarily and developed the quick frozen antihemophilic plasma

The realization that hematological progress could preserve life to an unexpected degree was followed by a new awareness of the extent to which hemophilia patients were disabled by crippling hemophilic arthropathies as they grew older At this point, the need for orthopaedic rehabilitation became apparent As open surgery and the customary methods for correction of deformities could not be employed because of the risk in hemophilia, it was natural to turn to a modality of treatment used successfully in the past in non-surgical correction of contractures and deformities caused by infantile paralysis Personal experience with the sound principle of subliminal forces introduced by F Mommsen in 1922 in a paper entitled "Die Dauerwirkung kleiner Krafte bei der Kontrakturbehandlung (Quengelmethode)" (Continuous Action of Small Forces in the Treatment of Contractures) suggested its adaptation to the special requirements of hemophiliacs

Treatment of our first hemophilia patient in 1946 for correction of a long-standing flexion deformity of the knee joint from hemophilic arthro-

pathy indicated the effectiveness of the method. Shortly thereafter, we launched a program for orthopaedic rehabilitation of patients crippled by hemophilic joint disease in cooperation with The National Hemophilia Foundation which offered to arrange for transportation of 25 selected patients from various parts of the country and to sponsor their treatment at Lenox Hill Hospital in New York City.

A diagnostic study of this first group showed a prevailing pattern of joint involvement and disability, and demonstrated without exception the feasibility of a method of treatment based on the original work by F. Mommsen. Practical procedures were soon established and, in May 1952, we were able to compi[illegible] a diagnostic and therapeutic survey of the first 25 patients correlating [illegible] and results of treatment in a scientific exhibit at the annual [illegible] of the New York State Medical Society.

After this publi[illegible] [illegible]x of hemophilia patients seeking orthopaedic rehabilitati[illegible] [illegible]luntary basis or supported by the Foundation reached th[illegible] more extensive studies based on larger series were pr[illegible] [illegible]merican Academy of Orthopaedic Surgeons in January 19[illegible] New York Academy of Medicine Graduate Fortnight on Bl[illegible] [illegible]n October 1953.

During this [illegible] National Hemophilia Foundation had grown considerably in [illegible] [illegible]d function and had decentralized its organization by esta[illegible] [illegible]hapters in key cities throughout the United States and Cana[illegible] [illegible]reatment of the hemophiliac at a local level could be handled [illegible]ore effectively and economically.

At the same time, orthopaedic surgeons interested in promoting local teams for treatment of hemophiliacs were invited to come to New York City to study at first hand the methods developed at Lenox Hill Hospital for correction of deformities and maintenance of rehabilitation by means of specially designed orthopaedic appliances.

Nevertheless, while local chapters of the Foundation were successful in organizing hemophilia families, and medical services became increasingly available with the progress in hematology, the orthopaedic aspects of hemophilia continued to attract painfully few practitioners in the fields of orthopaedic surgery and bracemaking. Even at the present time, The National Hemophilia Foundation and its New York chapter are depending largely upon the treatment center at Lenox Hill Hospital for rehabilitation of the more extensively crippled patients and particularly for their fitting with the individualized orthopaedic appliances developed for this purpose. Methods and results of our treatment during the past 10 years are published in the hope that they will demonstrate the possibility for human rehabilitation in this field and encourage the establishment of local facilities for orthopaedic treatment of hemophiliacs

Knowledge acquired in examination and diagnosis of 110 hemophiliacs

and extensive rehabilitory treatment of 98 of these patients between March 1946 and July 1956 forms the basis for this report. The original plan to recount case histories of the complete series of 110 patients was curtailed for financial reasons and detailed analyses were restricted to 56 Case histories for 35 of these are offered as abstracts while the remaining 21 cases, selected to illustrate particular or unusual problems and methods of treatment, are presented in full narrative form

Of the original 110 patients comprising this study, 60 are under observation and treatment as of January 1, 1958 Thirty-nine of these were part of the smaller study of 56 patients selected for extensive analysis and their case numbers are listed below for reference 3587 [illegible] 5112; 5630, 5637, 5653, 5682, 5733, 5737, 5747, 5926, 6003, 6131 [illegible] 6150, 6176, 6224, 6264, 6271, 6285, 6287, 6294, 6311, 6504, 651 [illegible] 6531; 6678, 6809, 6975, 7146, 7185, 7207, 7301, 7389, 7513, 75[illegible] [illegible]4, 8058.

While this report necessarily termin[illegible] et date of July 1, 1956, a considerable number of additio[illegible] ere examined and treated thereafter All of these gave furthe[illegible] the diagnostic criteria and effectiveness of methods of trea[illegible] ussed in these pages

As I am engaged in the private practic[illegible] ic surgery and in charge of the orthopaedic department [illegible] al, I could not have attempted the exacting treatment of [illegible] of patients and the evaluation of our methods without the co[illegible] istance of my co-workers, my hospital and The National Hemophilia Foundation I am therefore profoundly grateful to all the men and women who have supported this project and participated in the preparation of this book

The National Hemophilia Foundation, its founder and first president, Robert Lee Henry, and his wife, Betty Jane Henry, deserve much of the credit for inspiring the work accomplished to date in treatment of hemophilic arthropathies Without the efforts of Betty Jane and Robert Lee Henry to organize the Foundation and without their deep personal interest and constant encouragement, orthopaedic rehabilitation of hemophilia patients would never have developed to its present status in so short a time Supporting treatment of most of the earlier hemophilia patients at Lenox Hill Hospital, The National Hemophilia Foundation has continued to sponsor or at least act as a clearing house for hospitalization and medical care for hemophiliacs from all over the country

The Gustavus and Louisa Pfeiffer Research Foundation and Mrs Gertrude Sergievsky provided the funds for the preparation of this monograph Sincere thanks are due to these donors

As all this work was done at Lenox Hill Hospital, it is self-evident that I have depended upon the cooperation, advice and physical assistance of many members of the hospital Dr Francis H Ghiselin, Director of Radi-

ology, and his able staff of the X-ray Department have produced all x-ray pictures of our hemophilia patients They have expended a great deal of time and effort to provide superior films despite the difficult problems encountered in hemophilic joint disease. Dr Francis B Roth gave invaluable assistance in the treatment of the first hemophilia patients. He has also collaborated with me in compiling and displaying the scientific exhibits entitled "The Management of Hemophilic Arthropathies" which were presented in 1952, 1953 and again in 1957. Dr. Melvin Harris and more recently Dr. Karl R. Paley were responsible for the medical care of patients admitted to the hospital. Dr. Jacob Geiger, Director of the Lenox Hill Hospital Blood Bank, deserves credit for preparation, storage and availability of fresh blood and qu plasma for our hemophilia patients Dr. Alexander Levine has eat help with plaster of Paris work. He also gave much of h lection of x-ray films for this publication.

Mr Paul Schum er of the Lenox Hill Hospital Brace Department and a hig racemaker, has contributed most of the information relating ical procedures in the workshop. Mr. Schumacher merits re is part in the development of the hemophilia Hessing brace and paedic appliances as well as for the high caliber of the braces con ed under his supervision

In preparing this manusc pt for publication, Mrs. Carol S. Diamond has edited the entire text abstracted and written the narratives of the hemophilia case histories recorded in these pages. Devoting herself for many months to the cause of hemophilia, she has worked with excellent understanding of the medical, as well as the technical, details of the subject

A note of thanks is due to Miss Dorothy L. Goodenow who, for many years, has collected references from medical literature on the subject of hemophilia I am also indebted to Dr Walter R. Bett of London, England for his contribution to the bibliography

Mrs. Helen Vogel was kind enough to collaborate with me once again in preparation of the illustrations, and the Bergman Associates and Mr. Toni Ficalora have been responsible for the photographs. Mr. Ficalora, a former president of the New York Chapter of The National Hemophilia Foundation, generously donated the photographs he produced with such care for this publication

The manuscript was typed by my secretaries, Mrs. Elsa C. Brennan and Miss Linda Dubester

Finally, I should like to say a special word of thanks to the publisher with whom it was a pleasure and a privilege to work.

HENRY H. JORDAN, M.D.

New York, N Y.

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HEMOPHILIC ARTHROPATHIES

CHAPTER 1

Summary of Tabulations of 56 Patients with Hemophilic Arthropathy

I. FAMILY HISTORY OF HEMOPHILIA

TWENTY-EIGHT case histories contained no evidence of hemophilia or of any abnormal bleeding tendency in the present family unit or in the patient's ancestry In an additional 27 cases, patients (or their families) reported an incidence of hemophilia among one or more male relatives of the patient including grandfathers, great uncles of the patient's mother, great uncles, uncles, brothers, cousins, second cousins and, in one case, a hemophilic history of four generations on the mother's side The fifty-sixth case reported a bleeding tendency on the mother's part but no other occurrence of abnormal bleeding in the family Apparently, the patient's mother was reported to have bled considerably after a tonsillectomy and adenoidectomy in her youth, and profusely at the birth of the patient

II. CLINICAL ONSET OF HEMOPHILIA

The first indication of hemophilia among these 56 patients occurred in the form of subcutaneous, intramuscular, intra-articular or internal hemorrhages Some episodes followed surgery (including 14 at the time of circumcision) or major trauma, some were reported to be "spontaneous" *

During the first year of life	44
Age 1 to 5 years	8
Age 6 to 12 years	3
At college age	1

III. AGE OF PATIENT AT TIME OF FIRST ORTHOPAEDIC EXAMINATION †

Age 1 to 5 years	6
Age 6 to 10 years	17
Age 11 to 15 years	12

* There are no "spontaneous" hemorrhages, as all bleeding is initiated by trauma or microtrauma (movement of an extremity during sleep)

† Throughout the text, the term *first orthopaedic examination* or *consultation* refers to the first examination by the author

Age 16 to 20 years	5
Age 21 to 25 years	6
Age 26 to 35 years	5
Age 36 to 45 years	3
Age 46 to 54 years	2

IV. DEGREE OF DISABILITY AT PATIENT'S FIRST EXAMINATION

A *General disability.*

Total	22
Marked	25
Moderate	8
None	1

B *Functional disability for locomotion:*

Bedridden	21
Wheelchair ambulatory	8
Crutch ambulatory	18
Cane ambulatory	7
Ambulates with difficulty	2

V. OCCURRENCE OF HEMORRHAGES IN CYCLES

To date, no evidence has been established of cyclical behavior in hemophilia. Indications of the existence of "bleeding cycles" have been either the individual tendency toward variation in clotting time, or alleged bleeding patterns reported by patients or their families. For example, we discovered that a new patient who appeared to have a tendency toward excessive bleeding at the end of March had disregarded a large decubitus on his heel during the summer as there had been no bleeding from the wound Although the decubitus required considerable scraping of dead tissue, there was also no bleeding during this procedure. At other times, the patient would assuredly have bled profusely from such a wound. Another patient, finding summer months free of hemorrhages that afflicted him throughout the rest of the year, defined his hemophilia cycle as seasonal or climatic. Still another was almost free of symptoms when he spent the winter in Florida, and a fourth patient seemed more vulnerable when he lived in the hot climate of Houston, Texas These variations certainly appear to be less a characteristic of the patient's disease than an indication of environmental influence and cannot be termed cycles in the true sense It is known that clotting time varies within the individual; it is also accepted that certain factors such as climate and weather influence hemophilia as they affect other diseases. Discovery of the nature of their influence would perhaps lead to the development of protective measures that could be offered to hemophilia sufferers. Therefore, even superficial tendencies

or correlations presented by patients are of value in studying the relationship between hemophilia and external factors, and the following information is accordingly presented:

In 47 cases, no cycle or pattern was observed in the recurrence, onset or recession of hemorrhages In the other 9 cases, two reported that hemorrhages were most frequent and severe in the spring and fall, one was most afflicted by hemorrhages at the end of March; one had a much greater number of hemorrhages at the beginning of summer; one suffered major episodes in the spring and minor episodes in the fall, two were least troubled by bleeding during summer months, one experienced a complete bleeding cycle every 4 to 6 months, another hemorrhaged every 3 months, particularly in spring and fall.

VI. INTERNAL HEMORRHAGES

Among 56 patients, 32 reported having had internal hemorrhages before the first orthopaedic examination. Of these, 15 reported having additional internal hemorrhages after treatment was started, and 17 reported no further internal bleeding Of the 24 cases who had not suffered internal hemorrhages prior to inception of treatment, 14 continued to be free of them and 10 reported episodes of internal bleeding Distribution of internal hemorrhages was as follows·

Abdominal (including GI)	21
Urologic	21
ENT	12
Dental	7
Chest	4
Eye	3
CNS	3
Intracranial	2

VII. JOINT INVOLVEMENT

The following statistics deal with the first major joint affected in each patient, total number of major joints involved at time of first orthopaedic examination, total number of joints involved subsequent to first orthopaedic examination, and a summary of major joints affected among the 56 patients

A *First major joint involvement*

Knee	31
Elbow	4
Ankle	3
Hip	1
Shoulder	1
Multiple joint involvement	16

B. *Total number of major joints involved at time of first orthopaedic examination:*

Knees	88
Elbows	60
Ankles	36
Shoulders	10
Wrists	8
Hips	7
Total	209

At time of first orthopaedic examination, 7 of the 56 patients revealed involvement of only one major joint while 49 had multiple joint involvement. During span of observation, however, all patients eventually developed multiple joint involvement.

C. *Joints involved subsequent to first orthopaedic examination:*

Ankles	16
Elbows	12
Knees	9
Hips	9
Wrists	7
Shoulders	6
Total	59

D. *Total number of major joints affected among 56 patients:*

Knees	97
Elbows	72
Ankles	52
Shoulders	16
Hips	16
Wrists	15
Total	*268*

VIII. DEGREE OF JOINT INVOLVEMENT

Early hemophilic arthropathy	5
Moderate hemophilic arthropathy	9
Severe hemophilic arthropathy	38
Late hemophilic arthropathy	4

IX. PREVIOUS ORTHOPAEDIC TREATMENT

Exactly half of the 56 patients reported no orthopaedic treatment prior to first examination. Orthopaedic care for the other 28 cases included: aspiration and injections (Hyaluronidase, Hydrocortone), strappings, knee

caps, splints, various modalities of physical therapy, manipulation under anaesthesia, casts, traction, double-bar braces and foot plates.

X HOSPITALIZATION

For orthopaedic treatment of hemophilic arthropathies, 43 of the 56 patients required at least one hospital admission during the period of treatment, and 13 were treated completely ambulatory

A *Hospital admissions* Forty-three patients accounted for 88 Lenox Hill Hospital admissions totalling 1,506 days, of which 80 admissions totalling 1,337 days, were primarily for orthopaedic reasons, and 8 admissions, totalling 169 days, were for reasons not specifically related to bleeding into musculo-skeletal apparatus

B *Hospital admissions unrelated to joint bleeding*:

Tooth extractions	4
Abdominal hemorrhage	1
Gum bleeding	1
Intracranial hemorrhage	1
Kidney hemorrhage	1
Total	8

C *Concomitant problems* During hospital admissions for care of patients' hemophilic arthropathies, various complicating conditions existed and were treated concurrently. These included

1 Severe internal and muscular hemorrhages
2 Fracture of neck of femur
3 Malunited fracture of shaft of femur.
4 Fracture of tibia and fibula in healing stage
5 Reflex dystrophy of ankle and foot
6 Severe metatarsalgia from plantar callosities
7 Postpolio equinovarus deformity

XI ORTHOPAEDIC TREATMENT

A *Hospital treatment* During 80 hospital admissions for orthopaedic treatment of 43 patients, a total of 115 casts was applied, 70 plaster of Paris models were made, and 94 orthopaedic appliances were made and delivered (See Table I for distribution.) In 11 of these procedures, general anaesthesia was employed as follows

Avertin (rectal)	9
General (inhalation)	2

B *Additional ambulatory treatment* During periods of additional ambulatory treatment of these 43 patients, a total of 52 casts was applied, 82

TABLE I

ORTHOPAEDIC TREATMENT

	Quengel Casts	Wedge Casts	Maintenance Casts	Total Casts	Plaster of Paris Models	Hessing Braces	Fiberglass Braces	Foot Plates	Elastic Knee Braces	Splints	Double- or Spiral-Bar Braces	Total Appliances
Hospital treatment												
Upper extremities	3	–	1	4	6	–	6	–	–	7	–	13
Lower extremities	21	47	43	111	64	48	3	3	5	21	1	81
Total	24	47	44	115	70	48	9	3	5	28	1	94
Additional ambulatory treatment												
Upper extremities	–	–	4	4	12	–	12	–	–	12	–	24
Lower extremities	3	11	34	48	70	58	3	11	21	19	5	117
Total	3	11	38	52	82	58	15	11	21	31	5	141
Ambulatory treatment only												
Upper extremities	–	–	–	–	3	1	1	–	–	–	–	2
Lower extremities	1	1	8	10	37	24	2	9	10	9	4	58
Total	1	1	8	10	40	25	3	9	10	9	4	60
Total orthopaedic treatment												
Upper extremities	3	–	5	8	21	1	19	–	–	19	–	39
Lower extremities	25	59	85	169	171	130	8	23	36	49	10	256
Total	28	59	90	177	192	131	27	23	36	68	10	295

plaster of Paris models were made; and 141 appliances were made and delivered. (See Table I for distribution.)

C. *Ambulatory treatment only:* Ambulatory treatment only of the remaining 13 patients included 10 casts; 40 plaster of Paris models; and 60 orthopaedic appliances made and delivered (See Table I for distribution.)

D. *Total orthopaedic treatment:* Total orthopaedic treatment provided to 56 patients included 177 casts, 192 plaster of Paris models, and 295 orthopaedic appliances. (See Table I for distribution)

XII. DEGREE OF REHABILITATION

This section deals specifically with initial rehabilitation at the end of the first phase of active treatment, together with the time span required for this phase, as well as rehabilitation of stance and locomotion at the date of last contact with the patient.

A. *Twenty-one bedridden patients* Of 21 patients bedridden at time of first orthopaedic exemaination, 19 are now completely ambulatory with 1 or 2 appliances, and 2 are cane ambulatory with 1 or 2 appliances

Present Condition (21 Bedridden Patients)	*Rehabilitation at End of First Active Phase of Treatment*	*Time Span*
19 Amb w appliances	1 wheelchair amb	1 month
	1 amb between parallel bars	3 months
	1 cane amb	1 month
	2 crutch amb	1½ months
	3 amb	½ month
	7 amb	1 month
	4 amb	1½–3½ months
2 Cane amb w appliances	1 walking between parallel bars	2½ months
	1 cane amb	½ month

B *Eight wheelchair ambulatory patients·* Of the 8 patients who were wheelchair ambulatory at the time of first orthopaedic examination, 5 are now completely ambulatory with 1 or 2 appliances, 1 is cane ambulatory, 1 is currently under treatment for a recent hemorrhage, and 1, initially examined at age 43, preferred to remain wheelchair ambulatory and was therefore given only knee splints which eliminated pain at night As he declined treatment for severe hemophilic arthropathies at the knees, he is not included in the following tables nor in the statistics on page 11

Present Condition (7 Wheelchair Amb Patients)	*Rehabilitation at End of First Active Phase of Treatment*	*Time Span*
5 amb w appliances	2 amb	1 month
	1 amb	4 months
	1 cane amb	2 months
	1 crutch amb	9 months
1 cane amb w appliances	1 crutch amb	1 month
1 wheelchair amb	1 no change, still under treatment	1½ months

C *Eighteen crutch ambulatory patients:* Of 18 patients who were crutch ambulatory at the time of first examination, 3 are now ambulatory without appliances, 14 are ambulatory with 1 or 2 appliances; and 1 is now cane ambulatory.

Present Condition (18 Crutch Amb Patients)	*Rehabilitation at End of First Active Phase of Treatment*	*Time Span*
3 amb w/o appliances	1 cane amb 1 cane amb 1 amb w walker	½ month 1 month 1 month
14 amb w appliances	4 amb 5 amb 3 amb 1 cane amb 1 remained crutch amb	½ month or less 1 month 2–3½ months 1 month 1 month
1 cane amb	1 cane amb	1 month

D *Seven cane ambulatory patients:* Of these 7 patients, 2 are now ambulatory without appliances and 5 are completely ambulatory with one or more appliances.

Present Condition (7 Cane Amb Patients)	*Rehabilitation at End of First Active Phase of Treatment*	*Time Span*
2 amb w/o appliances	2 amb	½ month or less
5 amb w appliances	4 amb 1 amb	1 month or less 2 months

E *Two patients ambulatory with difficulty:* Both patients are now completely ambulatory with appliances.

Present Condition (2 Patients Amb w. Difficulty)	*Rehabilitation at End of First Active Phase of Treatment*	*Time Span*
2 amb w appliances	*2* amb w/o disability	1–3½ months

XIII. REDUCTION IN SIZE, TYPE OR NUMBER OF MAJOR ORTHOPAEDIC APPLIANCES

In 11 cases, correction of affected joints progressed to the point where the original need for major orthopaedic appliances was diminished or became non-existent as follows.

Number of Patients	*Original Orthopaedic Appliances*	*Present Orthopaedic Rehabilitation*
4	Ambulatory with 2 hemophilia Hessing braces	1 amb without appliances 1 amb without appliances, except for long trips 1 amb with HHB* and knee splint 1 amb with HHB and elastic knee brace

* HHB refers to hemophilia Hessing brace throughout text

3	Ambulatory with 1 hemophilia Hessing brace and 1 elastic knee brace	1 amb without appliances 1 discards HHB on alternate days 1 discarded HHB for spiral-bar brace
1	Ambulatory with 1 hemophilia Hessing brace and 1 spiral-bar lower-leg brace	1 discarded HHB for elastic knee brace except unusual activity
2	Ambulatory with 1 hemophilia Hessing brace	2 amb without appliances
1	Ambulatory with 1 elastic knee brace	1 amb without appliances

XIV. GENERAL REHABILITATION OF 55 PATIENTS

General rehabilitation refers to a patient's capacity to resume some degree of normal life after his disabilities have been treated and reduced so far as possible For a child or adolescent, general rehabilitation means a return to school and participation in less hazardous forms of play and sports, in adults, it covers the ability to work, travel, drive an automobile and enjoy a social life. Current records for 55 patients indicate the following

A. Present condition of 22 totally disabled patients at time of first examination

Complete rehabilitation with appliances	19
Cane ambulatory with appliances	3

B *Present condition of 24 patients with marked disability at time* of first examination:

Complete rehabilitation, appliances discarded	3
Complete rehabilitation with appliances	20
Under treatment for recurrent hemorrhages	1

C. Present condition of 8 patients with moderate disability at time of first examination·

Complete rehabilitation, appliances discarded	1
Complete rehabilitation with appliances	5
Under treatment	2

D Present condition of 1 patient with no significant disability at time of first examination.

No disability, appliance discarded	1

XV. DEATHS

During the period of observation, 3 of the 56 patients have died 2 from cerebral hemorrhages at ages 56 and 25, one from abdominal hemorrhages at age 23.

CHAPTER 2

Roentgenology

ROENTGEN EXAMINATION plays a significant role in diagnosis of hemophilic arthropathy and is even more important during orthopaedic treatment of the hemophilic patient. While original x-ray films of all diseased bones and joints, together with identical exposures of normal joints for comparison, form the basis for diagnostic classification of lesions, periodic Roentgen examinations of the patient are needed to indicate the best therapeutic approach and to check the progress of treatment.

TECHNIQUE

Roentgen examination of the hemophilic patient demands not only excellent technique from the photographic point of view, but also the observation of several precautions. The routine technique used at an x-ray department for extremity roentgenology must be adapted to the special requirements of the hemophiliac. Hemophilic extremities frequently show contractures and deformities which make it impossible to position the patient in the usual manner for A-P and lateral exposures. The most careful and sympathetic handling of the patient is employed to relieve anxiety and to prevent pain It is necessary to x-ray bones and joints in the position in which they are most comfortable The customary A-P exposure of a knee joint is almost useless if the knee cannot be fully extended. In most cases where flexion contractures at the knee are present, the so-called "tunnel view" is used to obtain the best information regarding the condition of the articulation. This "tunnel view" has become standard exposure for hemophilic knee joints

A special wooden appliance is used which holds the leg well supported with the knee in flexion of approximately 125 degrees. Whatever the degree of flexion at the knee, it is important to direct the center x-ray beam parallel to the tibial plateau Without this precaution, no proper information can be obtained regarding preservation of articular space.

Occasionally, flexible casettes are of value to conform to a particular deformity and to obtain pictures with the least possible distortion

A-P x-ray films of an elbow joint, which is rigid in flexion of 90 degrees or less, should be obtained by placing the elbow on the casette with the upper arm and the forearm at the same distance from the casette so that

the same angle is formed between the casette and both the upper arm and forearm If the center beam divides the angle of flexion at the elbow into *two equal parts, distortion of the articular surfaces will be minimized.*

Roentgen examination must always include pictures of the corresponding extremity, whether involved or not, to provide a basis for comparison of a pathological joint with a normal one, or of varying degrees of joint and bone involvement on both sides. These films must be taken under identical conditions All this requires more attention to detail and more time than needed for routine x-ray examination

Exposure data in hemophilia will vary a great deal from one individual to another because of the degree of decalcification of the osseous structures and increased soft tissue density from effusion and synovitis Once satisfactory x-ray films have been obtained, it is necessary to record the technical data For this purpose it is advisable to use a sticker on the x-ray envelope in which the apparatus used for the first Roentgen examination is recorded, together with information on film to focus distance, the KV, the milliampere seconds, whether screens have been used or not, whether a Bucky diaphragm was used, and, if possible, the identification of the technician Such technical data, recorded at the time of the first x-ray examination, will permit a repetition of identical exposures at the time of re-examination. Comparison of x-rays taken at intervals under identical technical conditions will facilitate the evaluation of the changes visualized in the new films

When it is necessary to take x-ray films of the hemophilic extremity through a plaster of Paris cast to determine the degree of correction achieved at a given time, the technician must keep in mind that plaster of Paris casts in the treatment of hemophilia patients are much more heavily padded than casts applied in treatment of fractures. Some experimenting with the technique of exposure will be required to yield films with sufficient contrast between cast and bones to permit recognition of structural details.

Roentgen examination is additionally important in the proper fitting and adjustment of orthopaedic appliances, especially for the weight-bearing lower extremities. Only true A-P and lateral exposures can reveal the relation of the anatomical axis of a knee or ankle joint to the axis of the hinges of a Quengel cast *X-ray films used to determine the fitting of orthopaedic* appliances, or the necessity for alteration of an appliance which has become too small, are taken with the patient standing and bearing full weight on the extremity. In these cases, true A-P exposure is preferred to lateral films Roentgen equipment must be adaptable to the height of the patient in order to direct the center beam exactly at the level of the axis to be checked (Figures 36 and 37).

DIAGNOSIS

The majority of our patients with hemophilic arthropathies are well known hemophiliacs. Roentgen examination will confirm the diagnosis. The significance of the x-ray picture, however, lies in the information which we can obtain regarding the degree of bone and joint involvement, the maturity of the skeleton at the time of the examination, the degree of deformity already present and the degree of additional deformity to be expected from further growth of the skeleton prior to maturity. The extent of generalized decalcification and the number and size of juxta-articular cysts will suggest the degree of softening of the bone. These factors will determine the plan for corrective treatment with plaster of Paris casts and orthopaedic appliances.

X-rays taken with the attention to detail essential in diagnosis and care of hemophilia patients will give a good picture of the preservation of articular space and of the destruction of articular surfaces This information, however, does not determine prognosis in hemophilic arthropathies as much as it might in other joint diseases Hemophilic joints that looked hopeless on the first x-ray films have been returned to surprisingly good function. Again and again, it is amazing to see how a marked degree of articular destruction with extensive thinning of cartilage leaving a joint space of only a few millimeters is compatible with a considerable range of motion.

Analysis of approximately 2,000 x-ray films of some 520 joints in 110 patients available for x-ray studies during a period of 10 years yielded extensive and recurrent information about hemophilic arthropathy, and x-ray follow-up of individual patients added greatly to our understanding of the nature of the hemophilic process. Consequently, certain recognizable patterns emerged and influenced diagnostic criteria, prognosis and treatment.

Our present classification of x-ray lesions of hemophilic arthropathy as early, moderate, severe and late is guided by the extent of bone and joint pathology and not by the involvement of one or several elements that make up the articulation.

The x-ray findings in acute hemarthros and in synovitis are non-specific and the picture is not characteristic for hemophilia. X-rays of an acute hemorrhage into the joint will show distension and increased shadow density of articular space, while synovitis without bone involvement shows thickening and increased density of the joint contour. No permanent x-ray changes were seen after single or recurrent bleeding into joint cavities. We have found that increased shadow density from capsular thickening and synovitis is usually transient and not a regular feature of the x-ray picture of hemophilic arthropathies In our x-ray material covering all age groups, we were surprised to find soft tissue thickening with increased shadow



Figure 1 Case #5624 (age, 2) Very early arthropathy, good correction of flexion contracture (bottom) but some posterior subluxation of *tibia*

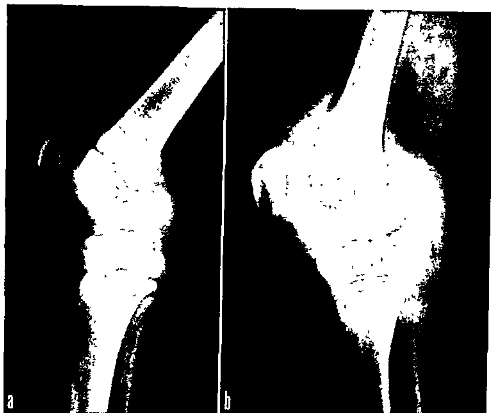


Figure 2. Case #7185 (age, 7). Left knee lateral. (a) 9/10/54, normal, (b) 6/18/56, acute hemarthros and first involvement of patella.

density absent in almost all cases of long-standing hemophilic arthropathy.

In contrast to these non-specific findings in hemarthrosis and synovitis, the *distinctive* findings in early, moderate, severe and late hemophilic arthropathy must be described.

In *early* hemophilic arthropathy we see small areas of cartilage erosion, irregular in contour and distribution, usually adjacent to subchondral defects and seldom visualized on opposing or contact articular surfaces. Epiphyseal changes may be present, consisting mostly of coarsening of trabecular structure with increasing decalcification There may, of course, be some shadow density from hemarthrosis and synovitis, either residual or recurrent. Many early cases show flexion contractures of the involved joints, sometimes with an early degree of subluxation of the tibia in relation to the long axis of the femur (Figures 1 and 2).

The picture of *moderate* hemophilic arthropathy is dominated by the presence of juxta-articular cysts of irregular distribution and size. These

cysts represent the most characteristic x-ray sign of hemophilia. Some of them show direct connection with the articular space because articular surfaces have broken down into these cysts with destruction of cartilage. This represents the first irreversible articular lesion

The epiphyseal changes seen in moderate as well as severe hemophilic arthropathy consist of side-to-side enlargement of the epiphysis, emphasized by narrowing of the diaphysis, thinning of cortex and decalcification of the osseous structures (Figures 3, 16a, and 17)

Ossification centers are premature in appearance due to accelerated growth, probably resulting from hyperemia. This is most frequently seen at the head of the radius, the patella, the medial condyle of the femur and the internal malleolus at the ankle (Figure 6).

Deformity of the patella, consisting first of over-all enlargement, particularly in its antero-posterior diameter, followed by squaring off of the inferior pole, is one of the most characteristic x-ray findings in hemophilia. Progressive cartilage destruction leading to thinning of articular space, as well as flattening of the articular contour, squaring off of femoral condyles or of the astragalus at the ankle are frequently seen

The *severe* case will show complete destruction of articular surfaces

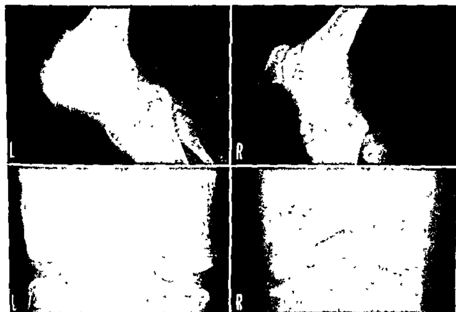


Figure 3 Case #5590 (age, 14) Left knee, very early hemophilic arthropathy, right knee, advanced hemophilic arthropathy with typical deformity of patella Tunnel view shows preservation of articular space bilaterally



Figure 4. Case #5759 (age, 13). Tangential exposures of patella right, early hemophilic arthropathy, left, normal.

and bizarre disorganization of metaphyseal trabecular structures unlike the x-ray picture of any other bone and joint disease (Figures 10, 11, 14, 16, 20 and 22).

In these three degrees of hemophilic arthropathy, the most important feature is hemorrhage into the bone, especially at the subchondral level.

Secondary changes are caused by faulty mechanics which lead to progressive cartilage destruction with thinning of joint space and flattening of articular contour

The severity of the disease is next determined by the degree of decalcification caused by bleeding into the bone and by a reflex mechanism from painful joint movements.

Hemorrhages prior to maturity of the skeleton invariably produce characteristic deformities These appear as side-to-side enlargement of epi-



Figure 5 Case #7207 (age, 7) Left knee lateral. (a) 9/19/54, (b) 6/18/56, good correction of flexion deformity but persistent posterior subluxation of tibia.

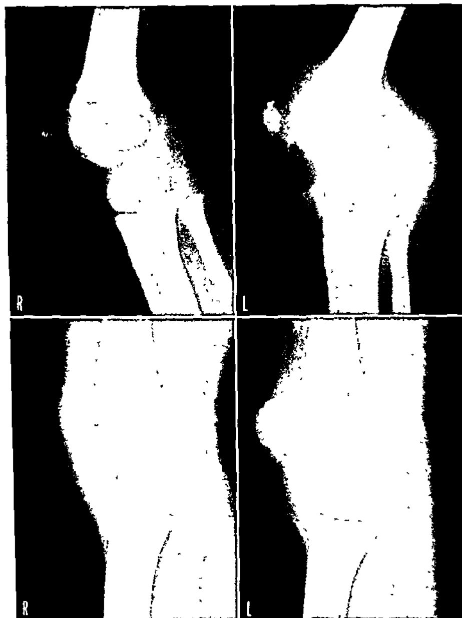


Figure 6 Case #7868 (age, 8) Right and left knee lateral (top) 9 21/55, (bottom) 8/16/56, overgrowth of patella of involved left knee, right knee normal



Figure 7. Case #6624 (age, 13). Right knee lateral (a) 12/10/53, moderate hemophilic arthropathy with marked deformity of patella, considerable decalcification, (b) 9/7/56, recalcification and retrabeculation under protected weight-bearing

physes and metaphyses emphasized by narrowing of diaphyses, thinning of cortex and decalcification, premature appearance and accelerated growth of ossification centers, premature closure of epiphyseal lines and increased length of the affected extremity.

Preponderance in growth of the medial femoral condyle over the lateral leads to genu valgum, aggravated in severe cases by an external rotation deformity of the tibia A similar mechanism causes valgus deformity at ankle and elbow.

If hemophilic arthropathy involves a knee joint prior to skeletal maturity, stimulation of growth will lead to early enlargement of the patella, followed by premature cessation of growth once hemorrhages have been stopped by adequate treatment. The distal third or apex of the patella is most affected. This leads to the characteristic picture of the "squared-off" patella found in almost every hemophiliac with knee involvement during childhood and adolescence. All stages of this peculiar development are well demonstrated in a 10-year x-ray study of a patient with only one knee affected, permitting comparison of growth of the pathologic and the normal patella (see Figure 9).

True ankylosis, or solid bony fusion, is apparently quite rare in hemophilia It was observed in only 3 of 449 joints (Figures 12 and 28).

In *late* hemophilic arthropathy, x-rays show secondary degenerative changes resulting from and superimposed upon the pathology of hemophilic joint disease which has long been inactive.

X-RAY INDICATIONS OF PROGRESS IN TREATMENT

Comparative x-ray studies of our patients will show the degree of correction of flexion contractures and other deformities as well as the extent of recalcification of osseous structures under protected weight-bearing It is gratifying to see how the trabeculation in the juxta-articular and metaphyseal zone becomes reorganized and presents a much more orderly though far from normal picture, than the bizarre appearance of complete disorganization of trabecular structure in a severely involved hemophilic joint prior to treatment (Figures 7, 10, and 11).

INCIDENTAL FINDINGS

While most of our Roentgen examinations were concerned with various degrees of hemophilic arthropathy and the results of treatment, our material shows a number of incidental findings which deserve some discussion.

Fractures occur in hemophilia patients just as they do in other persons Some of the fractures to be described are true fractures with the fully developed clinical picture of a fracture from severe trauma The x-ray studies of such a fracture of the femur with marked deformity due to over-riding and angulation of the fragments from inadequate immobilization until union had taken place, showed massive callus formation, a tendency to-

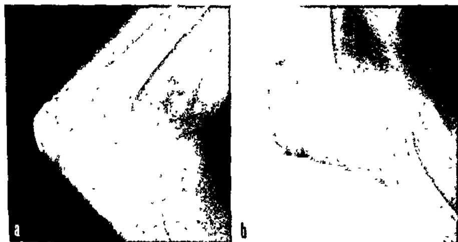


Figure 8 Case #7999 (age, 15) Left knee lateral (a) 1 7/56, flexion contracture of 115° Corrected by Quengel cast to 150°, under protected weight-bearing with hemophilia Hessing brace no more hemorrhages and good recalcification (b) 3 28/57, recurrence of flexion deformity with marked posterior subluxation of tibia after brace was outgrown and springs were broken



Figure 9 Case #3587 (age, 4, 1946). *Development of hemophilic patella:* (a) right knee 3/22/46, hemophilic arthropathy of two years' duration, (b) left knee 3/22/46, normal, (c) right knee 6/13/49, (d) left knee 6/13/49, (e) right knee 5/17/51, (f) left knee 5/17/51, still normal, (g) right knee 7/17/51, fully developed hemophilic *arthropathy with characteristic deformity of patella, good calcium density of osseous* structures With correction of flexion deformity and constant protection by cast and hemophilia Hessing brace since March 1946, no recurrence of hemorrhages into right knee (h) Left knee 7/17/51, early hemophilic arthropathy

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wards myositis ossificans, but also gradual decrease in the degree of deformity from angulation and over-riding as the patient grew (Figure 30).

In several cases a fracture was not detected because the trauma was either not reported by the patient or because the trauma was unusual In one case, a supracondylar fracture of the femur was discovered in the healing stage while the patient was undergoing treatment for a recurrent hemorrhage into the knee joint This fracture healed with solid bony union in a shorter than average time and the ensuing valgus deformity at the frac-

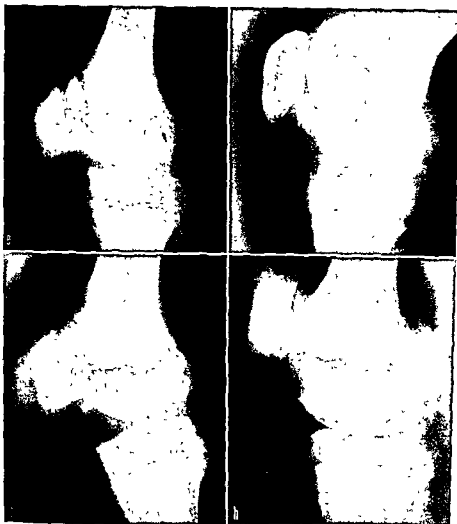


Figure 9 (*continued*)

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ture site corrected itself as the patient grew (Figure 31) In another case, a fracture through the proximal third of the tibia was also discovered healed but still recognizable after an injury which was not reported and which had not adversely affected an already present hemophilic arthropathy of the knee joint (Figure 32)

In a third instance, we found a regular fatigue fracture of the femoral neck in a 15-year-old boy who complained of pain in the hip but had suffered no hemorrhage (Figure 34)

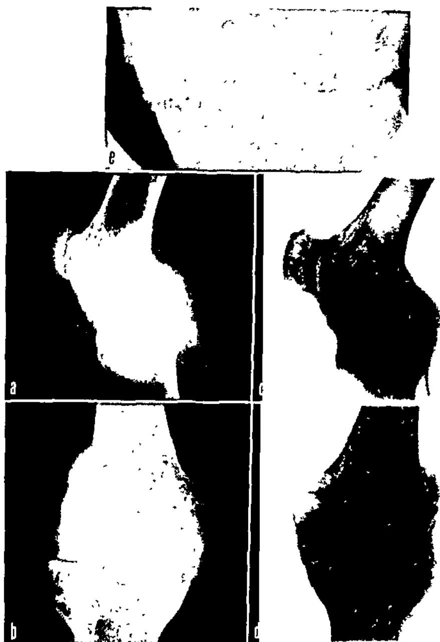


Figure 10. Case #5733 (age, 34). Right knee (a and b) Advanced hemophilic arthropathy Left knee (c and d) Advanced hemophilic arthropathy with large juxta-articular cysts including deformity of patella, (e) tunnel view demonstrates juxta-articular cysts, bizarre arrangement of trabeculations and slight preservation of articular space.

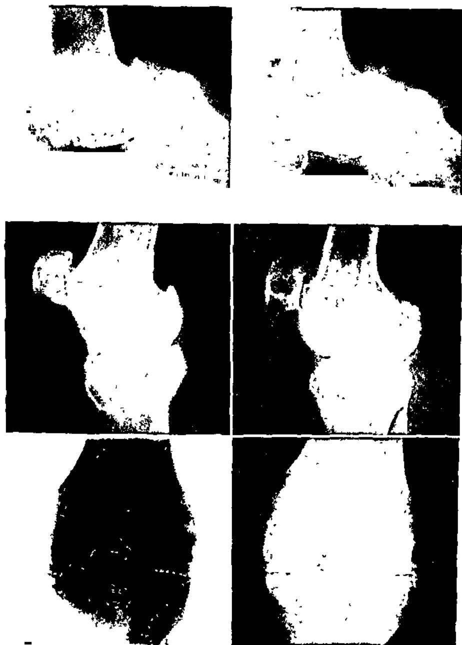


Figure 11 Case #5732 (age, 33) Advanced hemophilic arthropathy right and left knee, (top row) before correction of flexion deformity, (second and bottom rows) after correction of flexion deformity Many juxta-articular cysts, particularly large in patella *In spite of marked thinning of articular cartilage, good function and no pain on weight-bearing*

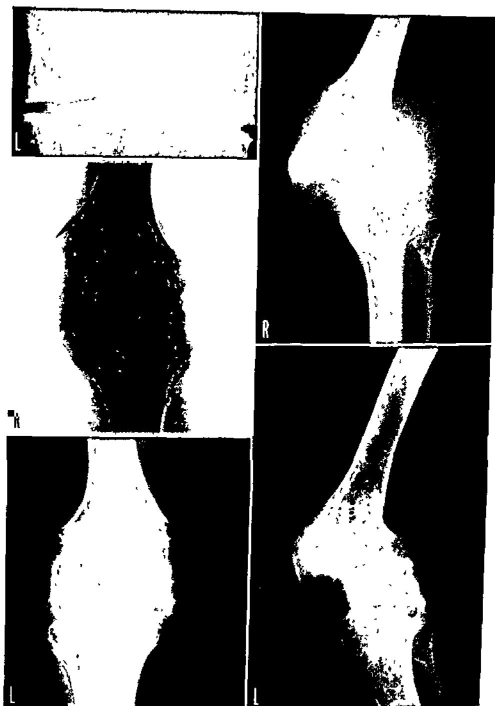


Figure 12 Case #5316 (age, 24) Right and left knee: advanced hemophilic arthropathy, true ankylosis right knee, typical deformity of patella, good recalcification of osseous structures under protected weight-bearing with hemophilia Hessing braces. Insert (top left) shows degree of preservation of articular space of left knee (tunnel view) in contrast to complete ankylosis of right knee.

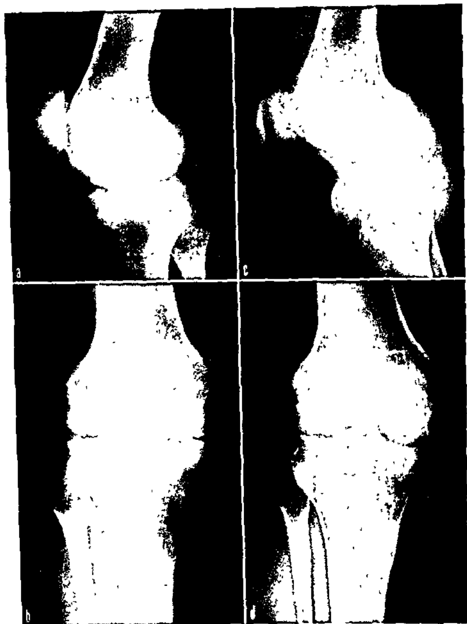


Figure 13 Case #5396 (age, 23) Left knee (a and b) 1/2/51, moderate degree of hemophilic arthropathy, (c and d) 6/4/51, after recurrent hemorrhage, considerable aggravation of articular involvement

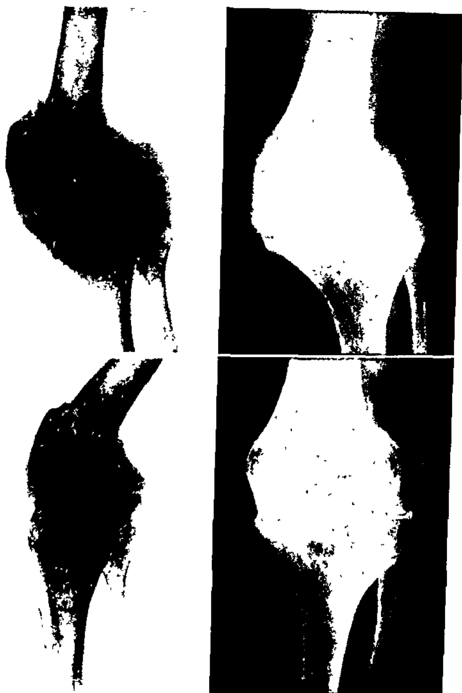


Figure 14 Case #8376 (age, 45) Advanced degree of hemophilic arthropathy right and left knee, (top) marked valgus deformity of more affected left knee

The worst fracture we have seen occurred in a knee joint affected by severe hemophilic arthropathy This patient suffered from epileptic seizures, resulting from a hemorrhage into the cranium earlier in life. One seizure occurred while he was sitting in an automobile on the way to the hospital for treatment of a hemophilic knee The muscle contraction of this particularly severe seizure forced the margin of the tibial plateau into the femoral condyles that were soft from decalcification and juxta-articular cysts An explosion-type fracture of the femoral condyles resulted with central separation and marked broadening of the distal end of the femur. Plaster of Paris immobilization followed by protected weight-bearing in a hemophilia Hessing brace, originally planned for the hemophilic arthropathy in the same knee, also promoted healing of the fracture with bony union and with much less permanent deformity than could have been anticipated at the time of our first examination of the boy (Figure 33).

One patient showed x-ray evidence of deformity and underdevelopment of forearm, wrist, hand and fingers due to Volkmann's ischemia from a severe crushing injury to the forearm and the wrist at the age of 18 months. The same patient had extensive involvement of the elbow of the same arm with unusually large juxta-articular cysts from hemophilic arthro-

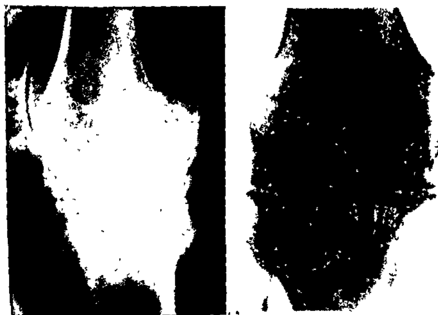


Figure 15 Case #6975 (age, 45) Advanced hemophilic arthropathy with good demonstration of massive juxta articular cysts and cysts in patella



Figure 16 Case #5630 (age, 6, 1951). Left elbow (a and b) 8/8/51, start of treatment, unusually large juxta-articular cysts in ulna, (c and d) 4/17/52, (e and f) 12/4/54, (g and h) 11/17/55 Right elbow (i and j) 12/4/54, normal, compare with (e and f) left elbow, same date Right elbow, (k and l) 11/17/55, with recent development of hemophilic arthropathy.

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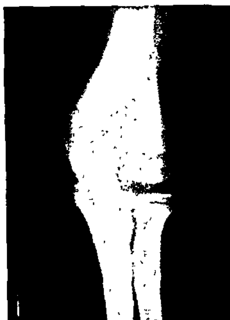
pathy The skeleton distal to the elbow showed no signs of hemorrhage into bone and joint in spite of the severe injury (Figure 16).

Finally, we have seen one case of microsomia, with underdevelopment of foot and ankle, from hemorrhage into the central nervous system (Figure 35)

Observation of these fractures in hemophilic patients indicates that fractures in hemophilia heal well and with solid bony union, usually in a shorter than average time for the patient's age. The speed of callus formation in hemophilia is probably due to the same influences that stimulate growth of affected epiphyses

DIFFERENTIAL DIAGNOSIS

As the majority of our patients were hemophiliacs with established diagnosis at the onset of treatment, there is little to be recorded in the way of differential diagnosis. Other states of decalcification must be borne in mind. Arthritis and degenerative joint disease, independent of hemophilia,


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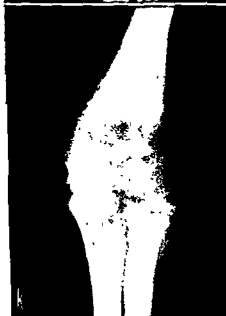

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Figure 17 Case #7207 (age, 7). Left elbow lateral: (a) 1954, early hemophilic arthropathy, (b) 6/18/56, progressive destruction

were observed. For instance, there were cases of degenerative joint disease from faulty static alignment and abnormal weight-bearing demands in an extremity which earlier in life had been the "good leg." None of the patients in this study exhibited x-ray evidence of gout, rheumatoid arthritis, bone or joint tuberculosis.

It is, of course, possible that a hemophiliac with arthropathies may sustain injuries to the musculo-skeletal apparatus without occurrence of hemorrhages. Thus we have seen a dislocation of a shoulder joint, as well as the above mentioned case of Volkmann's ischemia of the forearm from a severe injury during infancy.

The x-ray picture of a fully developed hemophilic arthropathy, especially if originating before skeletal maturity has occurred, is so characteristic *that it is sufficient to establish a diagnosis of hemophilia even if* certain clinical features, such as the clotting defect of the blood, are not yet confirmed.

Throughout the 10-year observation period on which this presentation is based, the ratio of distribution of joint involvement remained the same with the knee joint leading in frequency of involvement, followed by elbow, ankle and foot, while hemophilic arthropathies of wrist, shoulder and hip were less frequently encountered. The spine was never affected

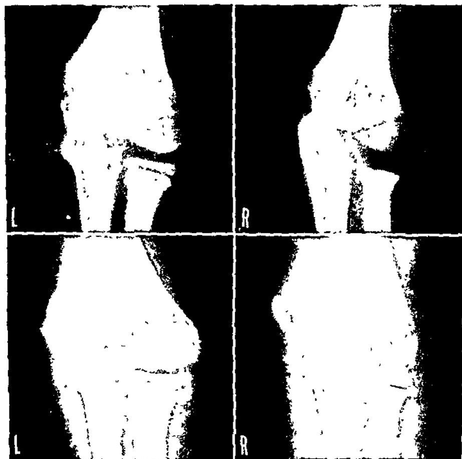


Figure 18 Case #7185 (age, 7). Right and left elbow A-P (top) 9/10/51, (bottom) 6/18/56, left elbow, moderate degree of hemophilic arthropathy, premature development of epiphysis and closure of epiphyseal lines, right elbow, normal



Figure 19. Case #5672 (age, 43) Left elbow A-P and lateral, former baseball player· involvement with hemophilic arthropathy after skeletal maturity, marked restriction of motion but no pain and no recurrent hemorrhages at this time of life.



Figure 20 Case #6975 (age, 45) Right elbow A-P and lateral: advanced hemophilic arthropathy with massive juxta-articular cysts.



Figure 21 Case #6271 (age, 16) Right foot and ankle, lateral (a) 1/17/53, ankle arthropathy and Sudeck's atrophy, (b) 1/14/54, recalcification of skeleton of right foot under protected weight bearing

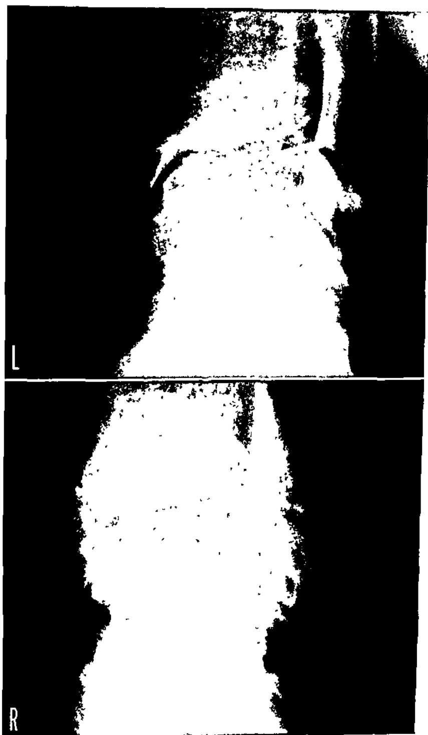


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Figure 23 Case #6376 (age, 45) Left shoulder (a) 9/28/56, long standing hemophilic arthropathy right shoulder (b) 9/28/56, more marked degree of hemophilic arthropathy, (c) 5/29/57 acute hemorrhage into right shoulder with marked distention of capsule, severe pain but no external signs of hemorrhage, recovery to previous state shown in (b) after two transfusions



Figure 24 Case #5272 (age, 6). Right and left wrist and hand A-P. Hemophilic involvement of left wrist and metacarpals with destruction of distal end of ulna, right, normal

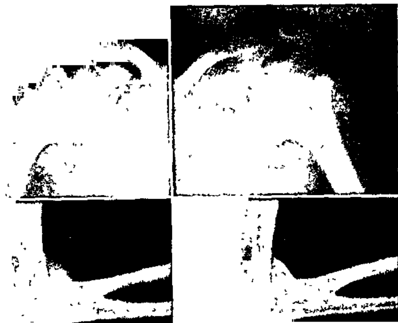


Figure 25 Case #5682 (age, 15) Figures 25-29 show patient with involvement of all extremity articulations deformity and disability of highest degree Right and left shoulder, elbow, wrist and hand are shown in Figure 25



Figure 26 Case #5682 (age, 15) Same patient as Figure 25 Pelvis and hip joints

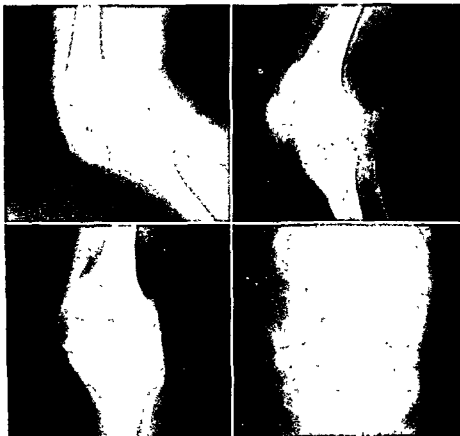


Figure 27 Case #5682 (age, 15) Same patient as Figures 25, 26 Right and left knee joints before correction of flexion contracture, marked ballooning of femoral condyles



Figure 28 Case #5682 (age, 15). *Same patient as Figures 25, 26, 27.* Right and left foot and ankle. True bony ankylosis of entire ankle and foot bilaterally.



Figure 29 Case #5682 (age, 15) Same patient as Figures 25, 26, 27, 28 Left shoulder (a) prior to crutch ambulation 10/4/51; (b) 10/13/52, shows deterioration of humeral head from use of crutch



Figure 30 Case #7513 (age, 9). Malunited shaft fracture left femur (a) 3/12/55, little callus, marked deformity, hemorrhages around the sharp end of the distal fragment, beginning calcification in soft tissues, (b) 5/24/55, massive callus formation and myositis ossificans, (c and d) lateral and A-P 10/16/56, healing of fracture, decrease in deformity under protected weight-bearing with hemophilia Hessing brace



Figure 31. Case #5737 (age, 11). Supracondylar fracture of left femur unrecognized despite definite trauma, treatment for hemophilic arthropathy of left knee. First x-ray films taken 3 weeks after injury showed healed supracondylar fracture with moderate valgus and flexion deformity and good callus formation. Later films showed complete disappearance of deformity.



Figure 32. Case #6271 (age, 16) Hemophilic arthropathy right knee, incidental finding of fracture of right tibia 1/17/53. Trauma 6 weeks earlier was not reported by patient. (*Right upper corner*) *right knee 9/25/56, complete healing of fracture. Left* knee, 1/17/53 hemophilic arthropathy originated earlier in life than that of right knee, hence marked deformity of patella Left knee now asymptomatic

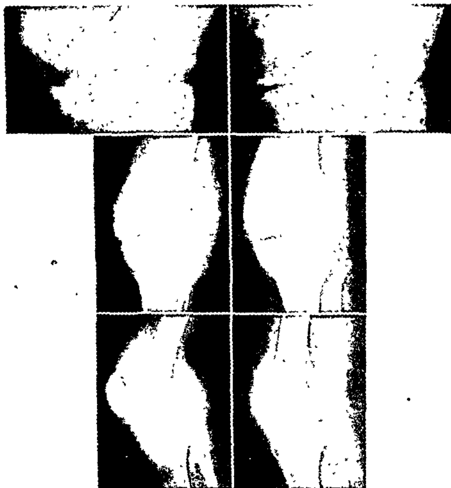


Figure 33. Case #7782 (age, 18). Right knee A-P, lateral and tunnel exposures. Explosive fracture of femoral condyle sustained during epileptic seizure (left side of picture), 8/29/55, fracture well healed without aggravation of pre-existing hemophilic arthropathy (right side of picture), 10/2/56.



Figure 34 Case #6224 (age, 14) Fatigue fracture right femur. No hemophilic involvement of right hip Hemophilic arthropathy right knee and ankle.



Figure 35. Case #8182 (age, 17). Microsomia left foot and ankle from hemophilic involvement of central nervous system. No hemorrhages into left foot and ankle.

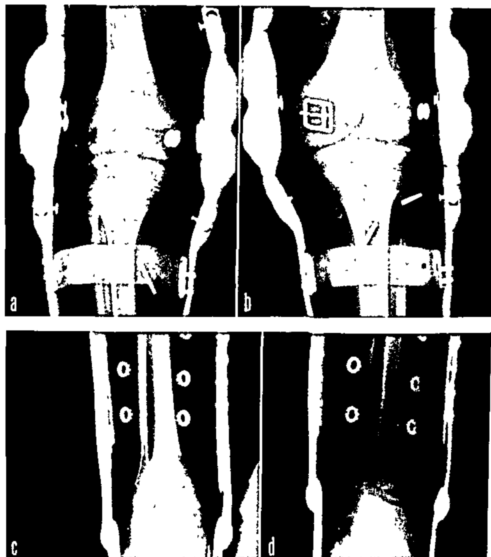


Figure 36 Case #3587 (age, 4, 1946). Same patient as Figure 9 (a and b) X-ray
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[illegible] ankle

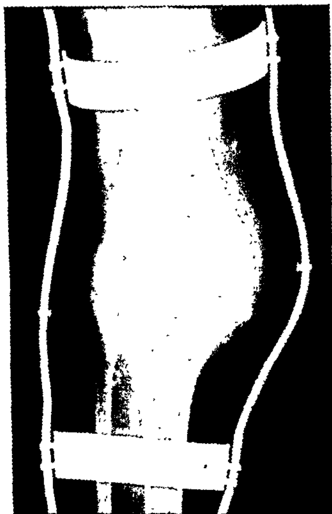


Figure 37 Case #8376 (age, 45) Same patient as in Figure 11 X-ray control of correction of marked valgus deformity from advanced hemophilic arthropathy of left knee in hemophilia Hessing brace constructed without knee joint

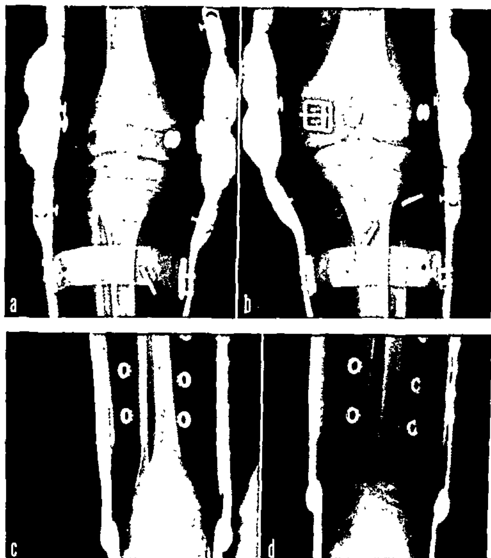


Figure 36. Case #3587 (age, 4, 1946). Same patient as Figure 9. (a and b) X-ray control of correct fitting of hemophilia Hessing brace for hemophilic arthropathy, right and left knees (c) X-ray check-up of brace fitting on right ankle, brace joint too low, *causing recurrent hemorrhages into ankle joint* (d) *Correct placement of brace*, ankle joint in line with anatomical axis.



Figure 87. Case #8376 (age, 45) Same patient as in Figure 14 X-ray control of correction of marked valgus deformity from advanced hemophilic arthropathy of left knee in hemophilia Hessing brace constructed without knee joint

CHAPTER 3

Plaster of Paris Technique

SAFE AND SUCCESSFUL orthopaedic rehabilitation of hemophilia patients demands the skill and judgment of the experienced orthopaedic surgeon. No phase of treatment in this field should be delegated to junior staff members or house staff. This is particularly true for the application of plaster of Paris casts which constitutes the basic and most important period in rehabilitation of hemophiliacs

Assuming that the reader is experienced in the use of plaster of Paris, discussion may be limited to the specific plaster of Paris technique developed to deal with hemophilia patients Materials and casts must be handled in prescribed ways to insure a consistently high level of performance

MATERIALS

Homogeneity of the plaster of Paris cast is prerequisite for strength and light weight Plaster of Paris is made from gypsum, a crystalline substance with the chemical formula $CaSO_4 \cdot 2H_2O$, by driving off the water of crystallization (about 21%). *When plaster of Paris bandages are immersed in* water, each molecule of $CaSO_4$ will take up the correct number of molecules of water to form crystals. Crystallization or setting takes place with a certain speed depending upon the quality of plaster of Paris used. As the homogeneity of the finished product can be disturbed by anything that will interfere with crystallization, instructions of the manufacturer should be carefully followed For instance, lumps of crystals within a plaster of Paris bandage due to exposure to humidity, partial wetting, or presence of foreign bodies of any type in the water from previous use, will form centers for faster crystallization Rubber gloves should be worn to permit removal of residual plaster of Paris on the hands by frequent rinsing. The water in the plaster of Paris *bucket should be changed after six 6-inch* bandages or an equal volume of 4-inch bandages have been used Temperature of the water should remain constant at approximately 103°F., unless otherwise specified by the manufacturer.

Setting time is also specified by the manufacturer. The plaster of Paris bandage should be held in a certain way to express all excess water over a "waste bucket" Inasmuch as the crystallization of plaster of Paris into

gypsum follows the reversible chemical equation mentioned above, the amount of water taken up by the calcium sulfate crystals is always the same. It is therefore advisable to express all excess water not absorbed during the specified immersion time.

These points, discussed in more detail in *Orthopedic Appliances,* Plaster of Paris Technique,* are particularly significant when handling hemophilia patients where atraumatic procedure and speed in the plaster room are of great importance.

Plaster of Paris Bandages

For plaster of Paris casts, 4-inch and 6-inch fast-setting or extra fast-setting plaster of Paris bandages of the highest quality are used. The fast-setting type, with a setting time of 5-to-8 minutes, is used for larger casts such as hip spicas and for casts in which hardware (i.e., hinges and posts) will be incorporated. Where good team work has been developed in the plaster room, bandages with the extra-fast setting time of 2-to-4 minutes may be used almost exclusively.

Plaster of Paris splints facilitate the quick application of a large cast or are used as reinforcements, particularly on the medial and lateral aspects of a wedge cast for the leg. Splints are made from 4-inch or 6-inch plaster of Paris bandages, rolled out dry to form a splint of the predetermined length. They are then folded in Leporello fashion, immersed, and expressed like a regular plaster of Paris bandage. Next, they are placed on a smooth surface, usually a board covered with linoleum, and all residual water is expressed while the plaster of Paris is rubbed smoothly into a homogeneous splint to be applied immediately. The setting time and the quality of the plaster of Paris used for splints must equal that of the bandages used in the same cast.

Padding

The following materials are used for padding:

Stockinette of various sizes
Sheet wadding
White felt of various thicknesses
Foam rubber
"Durocel" orthopaedic elastic padding bandage.
Crepe paper bandages
Wax paper

The possibility of individual hypersensitivity or even allergy to various

* Jordan, Henry H. *Orthopedic Appliances.* New York: Oxford University Press, 1939, Chapter II.

padding material must be considered to avoid irritation or itching of the skin and to eliminate the tendency of the patient toward scratching.

Stockinette should fit snugly without binding If it is too loose, it will form creases and folds causing pressure marks. The application of stockinette to the extremity is not possible in the presence of great pain from acute hemorrhage. The patient will not tolerate the pressure of rolling on the stockinette or the inevitable slight movements which occur when the stockinette is pulled on

Sheet wadding of finest quality may be used directly on the skin if stockinette cannot be applied.

Felt should not be used directly on the skin as it causes irritation in many patients. In addition to its usefulness as padding over pressure points and bony prominences, felt is also cut in strips 1-inch wide and placed beneath the plaster of Paris cast along the line to be cut for removal of the cast In hemophilia, it is advisable to wrap the strip in wax paper to prevent adherence of the felt to the plaster of Paris. This will make the use of the oscillating cast cutter safe.

Foam rubber sheets or *pads* are used over the sheet wadding to add resiliency to the padding usually at the points of application of the corrective forces of the three-point system to be discussed later in this chapter. The resiliency or recoil of foam rubber will not interfere with introduction of corrective forces at the right point

"Durocel" orthopaedic elastic padding bandage, recently introduced, has found a good place in casts for hemophilia patients. It cannot be substituted for sheet wadding or foam rubber but is valuable in maintenance casts where the padding may be of lesser thickness. Care must be taken to separate the "Durocel" padding from the plaster of Paris cast Originally, "Durocel" padding bandage was designed to form one layer with the plaster of Paris cast. For example, when the cast in a fracture case is bivalved, the "Durocel" padding remains firmly attached to the plaster of Paris and can be reapplied with the half cast This property of the "Durocel" bandage entails a risk in hemophilia The "Durocel" padding is cut with the cast and the cast cutter could penetrate too deep It is therefore necessary to separate the "Durocel" padding from the cast with a layer of crepe paper to avoid its adherence to the cast

Crepe paper bandages are used to hold the padding in place without danger of constriction as could be the case with gauze bandages They also separate the plaster of Paris from the padding, thereby promoting faster drying of the deeper layers of the cast by preventing the padding from absorbing some of the moisture of the wet plaster

Wax paper is used not only for wrapping the 1-inch felt strip along the

planned cutting line of the cast but also to separate layers of plaster of Paris which should come apart easily This is the case in a wedge cast where after each wedging for correction of a flexion deformity, a plaster cuff is applied over wax paper. At the next wedging it is then quite easy to make a longitudinal cut through the cuff down to the wax paper and remove the cuff in one piece.

Instruments

The oscillating Stryker electric cast cutter is the most important instrument Stille and other cast cutters of various designs are not suitable in hemophilia because their use entails too much levering and wrenching which would transmit some movement to the extremely sensitive hemophilic joint. The oscillating cast cutter permits the cast to be cut into any desired pattern without disturbing the part of the patient under treatment In hemophilia cases, the usual method of removing a cast by cutting it along the longitudinal midline, spreading it and bending it with duckbill and other instruments is generally inadequate as these procedures might cause too much movement and irritation of the joint Hemophilia casts should be cut into patterns, something like a jig-saw puzzle, so that each part of the cast can be lifted off the padding without changing the position of the limb.

Knives have to be handled with great caution and scissors, used for cutting the padding, should be very sharp but well protected

Special cast spreaders now on the market, with extra thin blades designed to conform to the cut made by the oscillating cast cutter, are superior to older types of cast spreaders for atraumatic handling of the patient

Hardware

Hardware to be incorporated into Quengel casts* consists of simple hinges, subluxation hinges, single aluminum posts for introduction of the Quengel, and double aluminum posts applied for simultaneous correction of both knee joints Turnbuckles of various types are needed for correction of adduction contractures at the hips Hinges with metal bridges to permit use of elastics for final extension of the knee joint are sometimes used All hardware to be incorporated into the cast should be made specially for the individual cast at the hospital's brace department

In addition to hardware, cord is needed for Quengel casts. The type used for window shades is most suitable for this purpose.

* See pages 63 to 70

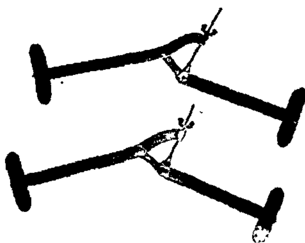


Figure 38 Subluxation hinges for Quengel cast.

Where elastics are introduced for minor or final correction of flexion contractures, No. 32 elastic bands are suggested.

Tongue depressors or pieces of aluminum or stainless steel of similar shape may be used as "shortsticks" in Quengel casts

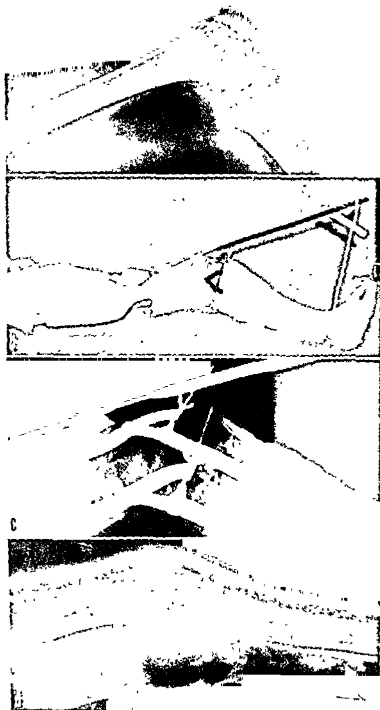
PLASTER OF PARIS CASTS

In the treatment of hemophilic arthropathies, the following plaster of Paris casts are used *protective casts, corrective casts, maintenance casts, special casts* and *negative models* or *molds* to be used in the manufacture of orthopaedic appliances

Protective Casts

Treatment of acute extremity hemorrhages employs the trias of plasma, immobilization and refrigeration. Protective casts have proved their usefulness in providing complete non-irritating immobilization of the extremity during the acute bleeding phase. Case after case in this series has demonstrated how wrong was the common belief that plaster of Paris casts could

Figure 39 Case #5637 (age, 12) Advanced hemophilic arthropathy right knee with typical deformity of patella and with posterior subluxation of tibia. (a) Lateral x-ray film with knee in maximum extension prior to correction (b) Quengel cast with subluxation hinges at the knee in action (c) X-ray control of progress of correction in Quengel cast (d) Maintenance cast applied after partial correction was obtained



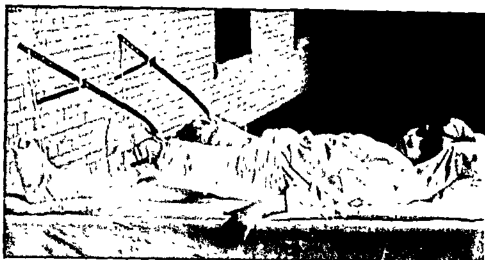


Figure 40 Case #5967 (age, 10). Quengel cast double hip spica for simultaneous correction of flexion contractures right and left knee in the presence of moderate hemophilic arthropathies No need for subluxation hinges

not be used during an acute hemorrhage into a joint, or into the soft tissues of an extremity.

When first seen, a patient is usually in poor general condition suffering from severe pain and lack of sleep. Analgesics and sedatives must be used immediately. While transfusion of whole fresh blood or plasma should not be delayed by application of a protective cast, it is frequently possible to have the transfusion running while the cast is being applied.

A protective cast aims to protect and immobilize the affected extremity without attempting the slightest correction of the flexion contracture or deformity which is usually present The most frequent condition needing a protective plaster of Paris cast is an acute hemorrhage into the knee joint or into the musculature of the calf The knee will be found in flexion of 90 degrees or less and the foot at times in marked equinus position Stockinette should not be used. Holding the leg in the position most comfortable for the patient, sheet wadding or "Durocel" is applied loosely and covered by large foam rubber pads Without exerting pressure, the whole padding is held in place with turns of crepe paper bandages. The plaster of Paris cast, extending from the toes to below the groin, is swiftly applied, without any attempt to mold the setting cast to any form or shape. As soon as the cast is rigid, the patient is returned to bed and placed in the position that is most comfortable.

A protective cast is quite bulky but this in no way reflects on the surgeon's plaster of Paris technique Its size and weight are unimportant as

the cast is only to be used during the acute phase when the patient is on complete bed rest. Such a cast may interfere with refrigeration. In some instances, it is advisable to cut a window of adequate size into the cast over the affected joint or muscle and to apply refrigeration with an icebag without cover, adding salt to the ice In other cases, the cast may be sufficiently short to permit refrigeration of the limb below the groin

After transfusion has stopped the hemorrhage, the patient will be com-



Figure 41 Hemophilic arthropathy right ankle Correction of equinus deformity by cast with turnbuckle-lever hinge

fortable regardless of the amount of swelling already present at the time of application of the cast. It has never been necessary to remove or bivalve the cast because it was not well tolerated.

After a few days, sometimes after twenty-four hours, the patient will report that the cast has become too loose and that he can move his leg within the cast The cast must then be changed and a new protective cast applied with the same precautions. It is usually found that some of the flexion deformity has disappeared, and that the new cast can be applied with the foot held in more dorsiflexion and the knee in greater extension. A smaller amount of padding is then permissible. After pain and recurrent bleeding have been eliminated, the patient is usually ready for application of a corrective cast

Corrective Casts

The majority of crippling hemophilic arthropathies concerns joints of the lower extremities, particularly the knee.

Rehabilitation of hemophiliacs affected in this way calls first of all for restoration of stance and locomotion After correction of deformities, many patients will have to rely upon special orthopaedic appliances for the lower extremities for periods of several years or longer. It must be emphasized, however, that an orthopaedic appliance is not in itself a suitable means of correction of a deformity. Orthopaedic appliances used in the treatment of hemophilia may introduce slight corrective forces to obtain a final or minor improvement beyond the point reached at the time the original orthopaedic appliance was fitted Plaster of Paris correction of flexion contractures and other deformities should always precede the use of an orthopaedic appliance

Preparation of Patient for Application of Corrective Casts

Reassurance of the patient is of great importance in avoiding emotional stress and its sequelae. The patient, if old enough to understand, will be more cooperative if he knows in advance every phase of treatment and its purpose Disappointment will be minimized if parent and patient can grasp the limit of achievement of each particular phase of treatment Wherever possible, contact with other hemophilia patients under corrective treatment or in various stages of rehabilitation is invaluable. It must be emphasized that the treatment will never cause pain. Pain would lead to reflex muscle spasm and defeat the purpose of treatment with "subliminal" corrective forces. The patient must realize that he will be handled with such care that there is little risk of hemorrhage. If a hemorrhage should occur coincidentally for other reasons, it can be readily controlled while he is in the hospital Sedatives may be used with discretion, while anal-

gesics should be strictly avoided during the period of corrective treatment. Under cover of analgesics, the patient might not experience and report pressure over bony prominences or pain in the soft tissues until too late He might also be too anxious to proceed with correction at a pace faster than that required for the soft tissues to readjust themselves to the new position of the skeleton.

Corrective orthopaedic treatment must be atraumatic in all its phases For this reason, it should not be necessary to use general anaesthesia except for an occasional patient who is extremely apprehensive *It may, however,* be used to assuage the discomfort of lying for a protracted period on the pelvic rest of a fracture table for application of a hip spica or double hip *spica with Quengel attachment*

Whenever anaesthesia is indicated, rectal Avertin is the preferred method Allowing for smooth recovery, this anaesthesia can be skillfully medicated to avoid excitation, retching, and straining, thereby avoiding hemorrhages into the abdominal wall, neck and throat The most experienced anaesthetist should be used and the procedure in the plaster room must be even more atraumatic than ever as the anaesthetized patient cannot report the warning signal of pain

Whether or not transfusion should be given before, during or after the treatment will depend upon the patient's general condition, particularly his clotting time Application of the corrective plaster of Paris Quengel or wedge cast should not produce even a microtrauma sufficient to start bleeding On the other hand, every precaution must be taken to avoid hemorrhages *independent* of the treatment which might interfere with the expected progress after application of the corrective cast

To avoid physical and emotional stress, the patient should spend as little *time as possible in the plaster room*

Atraumatic handling of the patient also calls for careful and rather slow motion throughout the initial application of padding To save time, it is necessary to be completely prepared with all materials needed for the application of a particular cast Measurements for hinges, subluxation hinges, metal bridges and Quengel posts must be made prior to application of the cast, and the hardware to be incorporated with the help of an experienced bracemaker must be ready and in good working order before the cast is started. Sufficient assistance and excellent team work are other prerequisites for swift and satisfactory plaster work in hemophilia cases

Two types of corrective casts, the *Quengel* cast and *the wedge* cast, have evolved as standard treatment for contractures and deformities Before discussing application of these casts, consideration should be given to the superiority of *a three-point system* of corrective forces over the traction method for the purpose of correcting flexion contractures

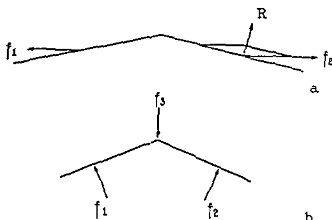


Diagram 1

Three-point System of Corrective Forces

Application of traction by ankle cuff, adhesive tape or plaster of Paris boot is not well tolerated and may be quite hazardous in hemophilia. The chief argument against traction, however, stems from mechanical considerations.

If traction is introduced to correct a flexion deformity of approximately 90 degrees, a parallelogram of forces will show that the resultant force for correction is *considerably smaller than the force of traction applied.* This is the case even if countertraction could be used efficiently. As the correction by means of traction proceeds and the angle of the deformity becomes larger or more obtuse, the resultant in the parallelogram of forces becomes smaller and smaller.

An entirely different situation is created when a three-point system of forces is introduced, with two forces opposing the center third force. In this case, the resultant force becomes greater with increased extension. A glance at diagram 1(a) and (b) shows immediately why a three-point system of corrective forces is more effective than traction

There is, however, another reason in favor of the three-point system, particularly for correction of *hemophilic flexion deformities of the knee* joint. In most of these contractures, a posterior subluxation of the tibia in relation to the long axis of the femur is present. Diagram 2 shows that the axis for flexion and extension of the knee joint changes in pathologic contractures from the normal axis of motion in the femoral condyles to a point between the articular surfaces of femur and tibia. A force applied for correction of the flexion deformity, especially when combined with posterior subluxation of the tibia, must avoid movement around the patho-

logic axis of movement as this would force the head of the tibia into the articular surface of the femur. Experience has shown that with softening of the bones from generalized decalcification and from juxta-articular cysts, the distal end of the femur is usually more affected than the harder and more resistant cortex of the tibial plateau

These principles of sound mechanics, based on the teachings of Hans von Baeyer, are pertinent to corrective plaster of Paris casts as well as to construction of all orthopaedic appliances

The Quengel Cast

The Quengel cast represents the best method of correcting flexion contractures of marked degree by introducing subliminal corrective forces which act continuously in the right direction and at the right point of a carefully planned three-point system of corrective forces Use of the Quengel cast necessitates complete bed rest and excellent nursing care, the procedure requires a great deal of patience on the part of the patient as well as the doctor. What can be accomplished with a properly applied Quengel cast, even in cases of marked deformity of long standing and in old and otherwise poor-risk patients, must be observed to be believed

The Quengel method was introduced by F Mommsen in 1922 in a

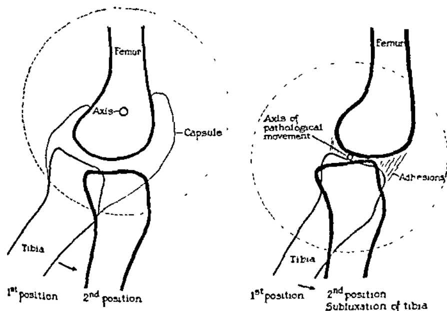


Diagram 2

paper entitled "Die Dauerwirkung kleiner Krafte bei der Kontrakturbehandlung (Quengelmethode)" ("Continuous Action of Small Forces in the Treatment of Contractures").

It was Mommsen's idea to correct the functional defect of a joint gradually over a long period of time, using minute forces which act constantly but which are not strong enough to cause pain and muscle spasm The chief obstacle to the correction of contractures consists in reflex spasm of the muscles involved. This may be overcome by the use of general anaesthesia as commonly employed in brisement forcé. But apart from the fact that forceful breaking or stretching of adhesions under general anaesthesia is not permissible with hemophilia patients, the general disadvantage of the brisement forcé is the trauma to which all the structures involved are subjected in order to obtain quickly the desired position. And while this position is maintained over a sufficient period of time to permit healing, new adhesions may form that can be the cause of another contracture. This in turn may necessitate a long and difficult course in physiotherapy, also undesirable in hemophilia. On the other hand, if the limb being treated is immobilized in a well-molded plaster of Paris cast to eliminate undue play, and if a corrective force acts uninterruptedly at the right place and in the right direction, the contracture will gradually give way and the soft tissue structures will rearrange themselves according to the new position

When planning the Quengel treatment, the contracture or deformity to be treated has to be thoroughly analyzed to find out the mechanical problem presented by each individual case. It is most important to determine the location of the axis around which the movement of the joint will have to take place and the interrelationship of the forces to be applied at this axis In calculating the extent of the problem, not only the natural axis of the joint and the position of the articular space must be considered, but also the fact that adhesions within the joint will change the mechanical relations of axis and levers once correction is attempted (Diagram 2, page 63).

Mommsen originated the use of a "Quengel" as the best means of applying a subliminal force. The so-called Quengel is a short-stick or toggle used for winding or twisting two cords, thereby gradually shortening their length. This Quengel had been most satisfactorily used in the treatment of contractures and deformities due to infantile paralysis in an era when surgery was not as freely employed as it is today Personal experience with the Quengel method and its modifications since 1922 suggested its adaptation for the treatment of deformities and contractures in hemophilic arthropathies in 1946 After application of 40 Quengel casts in treatment of 98 hemophilia patients, the Quengel is still considered the simplest and most

effective force and the one permitting closest control of corrective treatment.

The principle of the Quengel method is applicable equally to correction of flexion contractures of the upper and lower extremities and the plaster of Paris technique is much the same for all articulations lending themselves to this method. A Quengel cast is most frequently applied for correction of flexion contractures of the knee joint with restriction of extension to anywhere from 80 to 150 degrees, with or without posterior subluxation of the tibia.

Application of a Quengel Cast

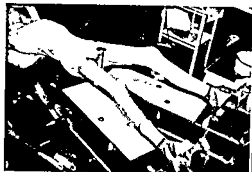
The accompanying illustrations show the application of a Quengel cast hip spica for correction of a flexion deformity of the left knee with posterior subluxation of the tibia in a 15-year-old boy

Flexion contractures of both knee joints had been previously corrected by the Quengel method and correction had been fully maintained for a considerable part of the sixteen-month interval between hospital admissions by *two* hemophilia Hessing braces The right knee joint remained asymptomatic despite the fact that the patient grew rapidly and was four inches taller than at the time his original orthopaedic appliances were delivered. The hemophilia Hessing brace for the left leg, however, was no longer effective Springs of the Lofstrand joints at the knee had broken and could not be replaced as there were no facilities for adequate repair at the patient's residence When he returned for readmission to the hospital, new x-rays showed considerable deterioration of the left knee joint with restriction of extension to 140 degrees and a very marked degree of posterior subluxation of the tibia Before this patient could be fitted with new orthopaedic appliances for both legs the flexion contracture of the left knee with posterior subluxation of the tibia required correction by the Quengel method

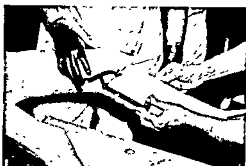
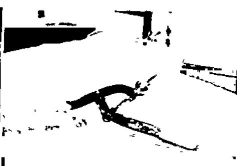
Prepared for treatment, the patient is placed on a table with pelvic rest, in this case on a Roger Anderson fracture table, with both feet attached to the foot sections of the table in the usual manner. A stockinette

as for incorporation of hardware are made prior to the start of the procedure to shorten the patient's time on the fracture table. The fastest-setting plaster of Paris (2-4 minutes) is used and, as far as possible, applied in splints.

The Quengel cast is applied in two sections the hip spica extending from the lower thorax apperture to the knee joint and the below-the-knee







[illegible] spica for flexion
[illegible] rent hemorrhages
[illegible] came ineffective

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section including lower-leg, ankle and foot. In applying the plaster of Paris, *the knee joint is held in a painless position without trying to correct any part of the deformity. Padding is applied from* the thoracic cage down to the ankle, while padding for the foot section has to be postponed until the foot can be released from the straps holding it to the fracture table. Points of maximum pressure during the correction, such as the dorsum of the thigh from patella to the groin, and the entire posterior aspect of the lower-leg are padded with foam rubber. This is covered with turns of sheet wadding. Ample sheet wadding must be used on the lateral aspect of the femoral condyles and particularly over the head of the fibula. A "dinner pad," which is a small sheet or two towels folded together, *is placed under the stockinette in the abdominal midline, to be withdrawn later for greater comfort when eating.*

When the padding is completed and held in place by turns of crepe paper bandages, the plaster of Paris cast is applied from the lower ribs down to the knee and, at the same time, from the knee to the ankle, leaving a small gap between the two casts at the knee joint.

As soon as the cast is rigid, the subluxation hinges are fitted by the bracemaker. These hinges are specially designed to permit simultaneous correction of the posterior subluxation of the tibia by tightening two wing *nuts on either side of the knee, while the Quengel itself works to straighten* out the flexion contracture.

Proper placement of hinges is of greatest importance for the desired result. First of all, there should be neither shearing forces nor incongruence between the hardware and the anatomical axis around which the corrective movement occurs. If simple hinges are used instead of subluxation hinges, as is permissible in cases where the subluxation is of minor importance or of very small degree, one can obtain some distraction at the knee joint and *anterior shift of the tibia during extension of the knee by placing the hinges* slightly anterior and distal to the anatomical knee joint axis which *is* at the level of the internal and external epicondyles of the femur.

In *this* case, subluxation hinges are used. The proximal joints of these

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(See Figure 8 for x-ray films.) (a) Patient on Roger Anderson fracture table, stockinette hip spica applied. (b) Application of felt and foam rubber padding to pelvis, dorsum of *thigh, posterior aspect of lower-leg.* (c) Completion of padding with sheet wadding held in place by crepe paper bandages. (d) Plaster of Paris hip spica is applied to left lower extremity excluding foot. Subluxation hinges and post for Quengel *are seen on right leg section of fracture table.* (e) The subluxation hinges *are carefully* fitted to correct axis position. (f) The subluxation hinges are incorporated. (g) The post for the Quengel is incorporated on dorsum of left knee. (h) Quengel in action.

hinges are placed exactly at the axis of knee joint motion, that is, at the level of the internal and external epicondyles which are palpable through the padding It is necessary to mark these two points for the proximal joints of the hinges on the cast with indelible pencil To obtain correct placement of the hardware, it is useful to use a tape measure to make sure that the thigh sections of the two metal hinges are at the same distance from the dorsal midline. In very difficult cases, it is advisable to determine with lateral x-ray films the correct site for the hinge using metal thumb tacks on either side of the cast. The distal joint of the subluxation hinge is used to bring the tibia forward while the knee is being extended The turnbuckle or wing nut screw is opened to give the maximum distance between the distal hinge of the appliance and the wing nut. Holding the hinge in this position, the lower-leg section of the hardware is placed in the long axis of the tibia and marked off on the cast with indelible pencil, again using a tape measure to insure equal distance from the dorsal midline for both hinges. The hinges must be parallel to each other to work in exactly the same plane. When adjusting with bending irons the thigh and lower-leg sections to conform to the contour of the cast, care must be taken to have sufficient distance between the hinges and the knee, as well as to keep them parallel in the sagittal plane. This can be accomplished by placing some plaster of Paris between the metal and the already rigid cast when incorporating the hinges by turns of plaster of Paris bandages. Once the location of the subluxation hinges in relation to the cast and to the knee joint is established, the most difficult phase of application of a Quengel cast has been accomplished

As soon as the hinges are secured, the aluminum post to which the Quengel will be attached is incorporated in the dorsal midline above the knee with the length of the post corresponding to the length of the leg below the knee. This means that a circle drawn around the center of the proximal hinge of the hardware would go through the heel of the foot as well as through the end of the aluminum post. When adding the aluminum post to the dorsum of the thigh section of the hip spica, it is advisable to place a few turns of plaster of Paris bandages around the proximal end of the post to insure better contact and more rigid incorporation into the already rigid hip spica.

Finally, the foot is released from the fracture table. Before this is done, the hip spica and particularly the knee section of the cast, must be supported in such a way that no force in the direction of extension is exerted on the knee joint The foot and ankle section is padded in exactly the same manner as the rest of the cast, using a foam rubber pad on the posterior aspect of the heel and throughout the sole of the foot, reaching slightly beyond the toes. After the padding is completed and held in place by crepe

paper bandages, the cast is finished by adding the foot section with the ankle held at approximately 90 degrees.

The stockinette at the proximal margin of the hip spica and at the toes is turned down and held in place with a few turns of plaster of Paris.

The cast is now complete and the patient is returned to bed with every precaution to prevent extension of the knee joint. After the cast is completely rigid and there is every evidence that it is well tolerated by the patient, a cord is placed around the heel section of the cast and connected to one of the holes drilled into the distal end of the aluminum post The type of cord used on window shades or Venetian blinds is most suitable for this purpose The cord is secured around the center of the heel section of the cast by a few turns of adhesive tape fastened onto itself to keep the cord from sliding The cord is tied to the post without undue tension

Correction starts by turning the wing nuts of the subluxation hinges at either side of the knee as tolerated Once the initial setting has been made, it is usually possible to turn the wing nuts one full turn each day, securing their position by small strips of adhesive tape About twenty-four hours after correction of the posterior subluxation of the tibia has been started, the Quengel or short-stick is placed between the cord close to the distal end of the aluminum post One or two tongue depressors are suitable for this purpose. By turning the short-stick or Quengel once in twenty-four hours for one-half or even for a full turn, as tolerated, correction of the flexion contracture is accomplished The tongue depressor is secured in place by strips of adhesive tape on the aluminum post as well as at the cord.

It takes almost longer to describe the technique of application of a Quengel cast hip spica than it takes to apply the cast With a good team of skilled technicians, the average time for a patient to remain on the fracture table may be reduced to thirty minutes.

When starting correction of the deformity by turning the wing nuts of the subluxation hinges and by tightening the cord with the Quengel tongue depressor, one must keep in mind that we are trying to correct a contracture the slow way by introducing "subliminal forces" only It would be easy to turn the Quengel more than once a day, and patients are frequently impatient and try to speed up correction by interfering with the carefully planned timing The cord of the Quengel should always appear quite relaxed This is difficult to convey to most patients, nurses and doctors The cord of the Quengel should never be taut as the strings of a violin Although the cord is relaxed, like a sail on a sailboat when no wind is blowing, it nevertheless accomplishes the correction almost miraculously, without causing pain, discomfort or the risk of a hemorrhage. This slow correction permits all soft tissues involved, for instance the vessels and

nerves in the popliteal region of the knee joint, to gradually adjust themselves to the changing position If the correction with Quengel and subluxation hinges is properly timed, there will be no setback. If the speed of correction is greater than the patient can tolerate, there will be a setback with valuable time lost.

The correction with the Quengel cast usually continues until the desired degree of extension at the knee joint has been obtained. This degree varies, of course, with the stage of treatment. For instance, if a Quengel cast is used for correction of a flexion contracture at the knee joint with extension limited to 80 or 90 degrees and with a marked degree of posterior subluxation of the tibia, one cannot continue with the first Quengel cast until extension of 160 degrees or more has been reached. This would take much too long and would entail too much physical and mental strain on the patient If we start to correct a flexion contracture of such marked degree, we will have to limit correction obtained by the first Quengel cast over a period not exceeding four weeks to an angle of extension of approximately 140 degrees At this time, the Quengel cast hip spica will have to be removed Carefully maintaining the angle of extension at the knee and avoiding any movement that would cause pain or muscle spasm, a plaster of Paris *maintenance cast* would then be applied from toes to below the groin With this cast, the patient may return to his home for a period of rest or even for a longer period during which he could go to school, returning at a later date to the hospital for further correction of the residual flexion contracture

In other cases, where it is evident that correction of the flexion contracture from a starting point of perhaps 120 degrees to a desired extension of 165 degrees can be accomplished within one three- to four-week period, the Quengel cast hip spica will be removed only when this degree of extension has been reached At this point, the leg is ready for a plaster of Paris model, to be used in the manufacture of a hemophilia Hessing brace with Lofstrand spring joints at the knee. The plaster of Paris model is made immediately after removal of the Quengel cast hip spica. Thereafter, without changing the position of the knee joint obtained by the Quengel correction period, a moderately padded maintenance cast is applied from the toes to below the groin maintaining the correction obtained to date. This maintenance cast will remain in place until the orthopaedic appliance is ready for fitting.

During the forty-eight hour period usually necessary to complete construction of the orthopaedic appliance after the fitting, the position of the knee joint can be maintained by reapplying the bivalved maintenance cast, or by applying a strong posterior felt splint with elastic bandages

The Wedge Cast

The wedge cast is much simpler to apply than a Quengel and does not require the help of a skilled bracemaker for incorporation of hardware. This is entirely a plaster of Paris technique. The procedure permits the patient to be wheelchair ambulatory and may serve to abbreviate the period of hospitalization.

The wedge cast is useful as the first and only cast for correction of moderate deformities, such as a flexion contracture of 150 degrees at the knee, a defect in dorsiflexion at the foot of only 10 or 15 degrees, or for correction of some types of contractures at elbow and wrist. Application of the cast must be very exact as it entails a greater risk of pressure over bony prominences, patella or heel, than does the Quengel cast. Nor is the position and movement of the joint under treatment controlled with the same precision as with a Quengel cast. The wedge cast technique changes the degree of extension of a joint at regular intervals, possibly once, twice or even three times a week, until necessary correction of the deformity has been achieved.

Application of a Wedge Cast

In describing the procedure for application of a wedge cast, attention will again be focused on the knee joint which is the articulation most frequently affected. The cast must extend from toes to the groin. It is not advisable to use a plaster of Paris cylinder from above the ankle to below the groin for this purpose because such a cast will eventually cause pressure and possible hemorrhage at its distal margin by riding down on the prominent external and internal malleolus. Moreover, the position of the foot without cast support is much less comfortable than if the foot is included in the cast with the ankle at 90 degrees. The cast should extend well beyond the toes on the sole of the foot and should end at the web between the toes on the dorsum.

The wedge cast may be applied over stockinette. Generous foam rubber padding should be used at the points of introduction of corrective forces.

→

Figure 43. Wedge cast for correction of residual flexion deformity right knee of 150. (a) The angle is measured and the cuts are outlined with indelible pencil. (b) The cuts are made leaving a bridge or hinge on lateral and medial aspects of the knee. (c) The dorsal wedge is removed. (d) Extension increased by 15°, correction maintained by insertion of small cork. (e) Cast closed by circular plaster of Paris over wax paper to facilitate separation of cuff from basic cast at next wedging of cast. (f) The cuff is removed for additional correction, note wax paper inside of plaster cuff.

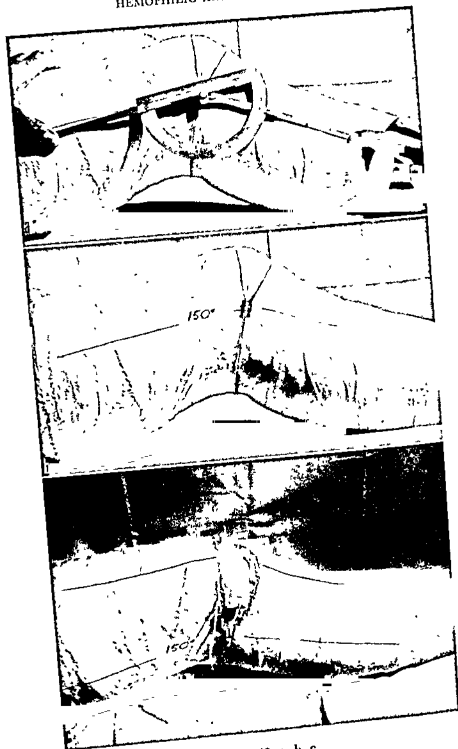


Figure 43. a, b, c

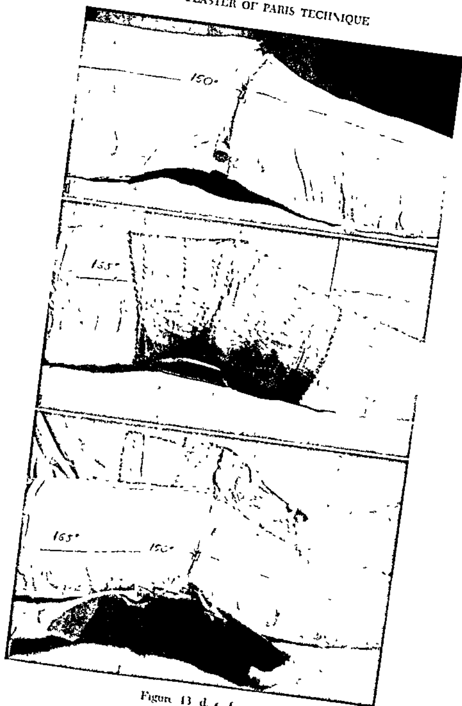


Figure 13 d, e, f

particularly throughout the dorsum of the thigh down to the apex of the patella and on the posterior aspect of the lower-leg from the popliteal region to the heel and sole of the foot. The foam rubber padding is held in place by circular turns of padding with sheet wadding and finally by crepe paper bandages. During application of this cast, the knee is held comfortably extended and great care must be taken not to introduce the slightest force in the direction of extension.

To facilitate later removal of the cast, a 1-inch strip of white felt within a sheath of wax paper is applied in the dorsal midline from one end of the cast to the other. This, too, is held in place with a few turns of crepe paper bandages. *The cast is then applied in the usual manner, using circular* turns, reinforced by plaster of Paris slabs on the medial and lateral sides of the leg but not on the anterior or posterior aspects where the wedging takes place. The sole of the cast may also be reinforced with a plaster of Paris slab although a wedge cast is not intended as a walking cast

The wedging process for correction of a deformity cannot begin unless the cast is completely comfortable, causing no pain or pressure at any point. To make sure that the cast is well tolerated, at least twenty-four hours must elapse after the cast is rigid and trimmed before the first wedging can take place.

Wedging of a cast involves removing a dorsal section at the level of correction, that is, on the dorsum of the knee joint, and cutting the cast in a straight transverse line on the posterior aspect in the popliteal region *This leaves a plaster of Paris hinge or bridge about 1-inch wide on* both sides. The position of this bridge must be exactly at the level of the axis around which movement of the knee joint during the corrective procedure will take place This is usually at the level of the medial and lateral epicondyle of the femur.

To obtain correct placement of the bridges on both sides of the cast, to remove the dorsal wedge and to cut the cast posteriorly at the right place, it is necessary to make an indelible pencil drawing on the cast.

First, the dorsal midline over the patella is determined and marked. The circumference of the cast at this level is then measured and divided by four One quarter of the circumference is now traced *from the dorsal* midline down to the region of the medial and lateral epicondyle of the femur. The axis of the movement intended by wedging is thus determined *At this point,* a bridge of 1 inch, half above and half below the axis, is marked off. This bridge is not to be cut because it will serve as a hinge for the motion of the knee Using a flexible tape measure, the proximal margins of the plaster of Paris bridge on either side of the knee are now connected by two lines, one running along the proximal border of the

patella, the other below the distal pole of the patella. Thus a wedge of elliptical shape, similar to the slice of an orange, is traced This section of the cast is then carefully cut out with the Stryker electric cast cutter Thereafter, the patient is turned into prone position with the foot section of the cast well supported by sandbags The distal margins of the plaster of Paris bridge, marked off on either side of the cast, are connected by a straight line and the cast is cut along this line Very minute pressure is used to extend the knee joint A plaster spreader may be inserted into the cut at the posterior aspect of the cast Extension should not be forced to the point where the patient complains of discomfort or pain It is usually possible to correct at least 5 degrees at one session A small cork is placed into the gap in the popliteal region of the cast to maintain the degree of correction while the cast is closed As correction progresses and the knee becomes more extended, a larger cork will be used Care must be taken not to break the plaster "hinges" at either side of the cast. The patient is turned back into supine position with the cork in the popliteal region maintaining the position of extension obtained at this wedging To facilitate further wedging, the center of the cast is wrapped in wax paper for several inches above and below the knee This section is now covered with circular turns of a 6-inch plaster of Paris bandage, securing the position until the next wedging takes place

For the second wedging, it is only necessary to cut the circular plaster of Paris in the dorsal midline down to the wax paper which facilitates separation of the cuff from the basic cast. After additional five degrees of extension have been obtained, a plaster of Paris cuff is applied over wax paper.

The wedging procedure can be repeated at regular intervals until the desired degree of extension, usually 165 degrees, has been reached At this point, the patient is ready for a plaster of Paris model of his leg, to be used in the manufacture of a hemophilia Hessing brace This is followed by application of a maintenance cast which is worn until the orthopaedic appliance can be fitted

Maintenance Casts

Fatigue and emotional strain, often the result of prolonged hospitalization and treatment, may cause an increase in clotting time and hemorrhages It is therefore essential to limit active treatment to periods not exceeding one month. Maintenance casts are used during these rest periods to maintain the correction achieved by Quengel or wedge casts

Great care must be taken not to irritate the joint during the change from corrective to maintenance cast While maintaining the degree of flexion or

extension of the joint under treatment, no attempt should be made at further correction while applying the maintenance cast. The cast for the lower extremity will usually extend from the toes to below the groin Plaster of Paris cylinders starting above the ankle are not satisfactory maintenance casts as they tend to slip down and cause pressure above the ankle or over the patella no matter how well they are applied.

The maintenance cast should be stronger than corrective or protective casts because the patient may now increase his activities Padding may be limited to stockinette, sheet wadding or "Durocel" with a strip of felt in a wax paper sheath applied to the dorsal midline or to both sides of the extremity to facilitate removal of the cast.

With such a cast, some patients may be allowed crutch or cane ambulation, while others have to remain wheelchair ambulatory. The maintenance cast may stay in place for one, two or three months depending upon the original plan for treatment.

Special Casts

Special casts were required in a minority of cases as the basis for corrective or elastic splints for hands and fingers; in connection with a partial orthopaedic appliance for an extremity; or, where correction of flexion deformities at both hips, as well as flexion contractures of both knees, called for a very large, heavy and cumbersome cast. In one case of multiple joint involvement, the weight of the cast alone would have caused pressure and probably bruises and hemorrhages in a patient with a long clotting time It was necessary to place this patient with large double hip spica in balanced traction to the overhead frame of a fracture bed, thereby eliminating the risk of too much pressure At the same time, gravity was utilized instead of a Quengel for correction of the flexion contracture at both hips (see Figure 62d)

Plaster of Paris Negative Models or Molds for Orthopaedic Appliances

After satisfactory correction of a deformity or flexion contracture has been achieved by the use of corrective Quengel or wedge casts, the patient is ready for fitting with an orthopaedic appliance.

All major orthopaedic appliances should be made to plaster of Paris models

During application of the plaster of Paris cast to be used as a mold for a positive model on which the orthopaedic appliance is designed, the same atraumatic handling of the patient is required as for all other phases of treatment. Joint irritation leading to hemorrhage must be avoided At the same time, care must be taken not to lose any degree of correction already ob-

tained. Whenever a plaster of Paris model is made, the joint or extremity under treatment must be quiescent, that is, without pain, swelling, redness of the skin or local heat. The plaster of Paris cast is applied over one layer of stockinette. *To facilitate removal of the cast, a 1-inch strip of felt in a* wax paper sheath is placed along the longitudinal midline and, if necessary, held there with crepe paper. Bony landmarks and joint axes are marked on the stockinette with indelible pencil. The plaster of Paris is then applied as fast as possible and this time carefully molded to the bony prominences. When the *cast is almost rigid,* it is cross marked with indelible pencil along the cutting line to secure adaptation of the cut edges of the mold after removal. After cutting the cast over the strip of felt, this strip is pulled out and the stockinette is slit with bandage scissors Using as many assistants as are available, the cast is slowly and carefully spread apart sufficiently to lift the limb from it Considerable help is required to maintain unchanged the position of the extremity after the cast has been removed, while the bracemaker and his assistant close the cast to the mold by turns of wet gauze bandages

The making of this mold of the extremity is followed by reapplication of a maintenance cast which may be light in weight as it will be worn only for the week or ten days until the orthopaedic appliance is ready for fitting

CHAPTER 4

Orthopaedic Appliances

THE SECOND important phase in rehabilitation of the patient crippled by hemophilic arthropathies covers fitting with orthopaedic appliances to maintain the benefits achieved under the corrective treatment discussed in the previous chapter. The history of hemophilia patients will, in most cases, demonstrate that enduring rehabilitation relates in large measure to the quality of the orthopaedic appliance.

Unfortunately, the level of bracemaking throughout the country does not meet the high standards required for the treatment of hemophilia sufferers Efforts of The National Hemophilia Foundation to organize hemophiliacs on a local basis and to decentralize hematological treatment by creating chapters throughout the country have been highly successful But there has been a tremendous lag on the orthopaedic side This was partly because it was difficult to interest a sufficiently large number of orthopaedic surgeons in the treatment of hemophiliacs, but primarily because there were only limited facilities for design, construction, fitting and repair of the individualized type of orthopaedic appliance required by the hemophilia patient As a result, many of these patients still have to make long and expensive journeys to New York City for orthopaedic treatment as well as for adjustment and refitting of their appliances at regular intervals.

For the hemophiliac has no alternative to the orthopaedic appliance that must follow corrective treatment As long as he cannot submit to surgery for correction of deformities, stabilization of joints, or elimination of severely diseased and painful articulations by arthrodesis, he must depend upon highly individualized brace construction for permanent rehabilitation.

Bracemaking, the oldest branch of orthopaedics, has always been and still remains an art, attracting too few of the sorely needed technicians to its exacting and sometimes tedious methods. Even with ever-increasing progress in mechanical fields, bracemaking continues to depend largely upon the craftsmanship and experience of those concerned with designing and building orthopaedic appliances.

It is true that new plastic materials such as fiberglass, and metals with greater strength and lighter weight, including aluminum and magnesium, are being utilized in the manufacture of these appliances But the fact remains that an individual brace, planned for a person with a specific de-

formity, depends upon the skill with which a mold of the extremity is made for a plaster of Paris model and the technique employed in handling the various materials to their best advantage in brace construction.

We have always emphasized that individual brace construction to plaster of Paris models must be superior in every instance to a brace assembled from stock parts. This is particularly true for the hemophilia patient who depends more upon individualized brace construction than patients crippled by infantile paralysis, cerebral palsy, arthritis or malunited fractures.

As has been stated, correction of deformities by Quengel and wedge casts will rarely match the results obtainable in non-hemophilia patients by the use of surgery. The maximum benefit that can be derived from corrective plaster of Paris treatment is a fair weight-bearing alignment. With hemophilic arthropathies, factors such as accelerated growth of the medial femoral condyle and the internal malleolus will cause some degree of valgus at the knee, external rotation of the lower-leg, valgus position of foot and sometimes restriction of dorsiflexion at the ankle. There may often be a considerable degree of residual posterior subluxation of the tibia in relation to the long axis of the femur, thus making the decalcified and large femoral condyles quite prominent. Aside from being more difficult and more hazardous to fit stock parts to these deformities with bending irons, it is less accurate than to start at the beginning by taking a plaster mold of the extremity to be braced. As the hemophiliac is vulnerable to the slightest trauma, undue pressure may cause subcutaneous or intramuscular bleeding. An ill-fitting brace can easily produce a decubitus, which could be catastrophic for a hemophiliac. In addition, incongruence of brace and anatomical axes will create shearing forces which, in hemophilia, lead to hemorrhages into joints. Furthermore, if the commonly used double-bar leg braces were sufficiently padded to protect the vulnerable hemophiliac, much of the needed support would be lost through the resiliency of the padding.

The orthopaedic appliance for the hemophiliac must be designed to spread over a very large surface of the body the forces required for maintenance of correct weight-bearing alignment in order to reduce the pressure per square inch of the extremity. This is even more important if Lofstrand spring joints or elastics are used for additional correction. Such large surface contact between brace and extremity is best accomplished by the use of leather, that is molded over a plaster of Paris model. In addition to protecting the patient against undue and uneven pressure, the appliance must prevent harmful motion or motion in the wrong direction.

In some cases, a patient's occupation and type of disability will demand that the orthopaedic appliance include a means for restriction of motion. For instance, if an ankle joint with restriction of dorsiflexion to 100 degrees

is braced, the orthopaedic appliance must introduce a stop to prevent the foot reaching the limit of dorsiflexion of 100 degrees. The reason here is that if pressure is exerted on the foot while the patient is driving a car, and dorsiflexion of the brace is not limited, the articular surfaces diseased by hemophilic arthropathy would be subjected to constant trauma leading to new hemorrhages

While providing protection, stability in the best possible weight-bearing alignment and axis-correct motion for movable joints, the brace may also introduce further subliminal forces by means of Lofstrand spring joints or added elastics to obtain greater extension of a joint than was present at the end of the plaster of Paris period.

All these requirements of the hemophilia patient can be fulfilled only by an orthopaedic appliance made to an accurate plaster of Paris model, constructed scientifically from the best materials, fitted without additional trauma and finished with particular consideration so that the orthopaedic appliance itself should not entail a risk at any point.

Why is it so difficult to find adequate facilities for this type of brace construction?

One reason is that the number of hemophilia sufferers in need of orthopaedic appliances is, of course, infinitely smaller than the large section of the population requiring appliances for the sequelae of infantile paralysis, cerebral palsy, malunited fractures, hemiplegia and arthritis.

Orthopaedic treatment for these patients, strongly influenced by The National Foundation for Infantile Paralysis, has successfully used various types of braces that can be readily assembled from stock parts. In our highly mechanized age, great effort has been expended to improve the materials and construction of assembly braces. They entail much less labor and are considerably less expensive than, for instance, the individualized molded leather-steel hemophilia Hessing brace. And, for most of the patients mentioned, they are satisfactory. As a result, there are too few brace shops interested in and capable of producing the individualized brace needed for the rehabilitation of the hemophilia patient.

In outlining the most felicitous conditions for individualized bracemaking, cooperation between orthopaedic surgeon and bracemaker cannot be overemphasized A brace shop within the confines of the hospital, and supervised by the surgeon, would be the ideal arrangement toward this end. While the surgeon must educate the bracemaker to the special needs of the hemophilia patient, he, in turn, must be familiar with materials used and techniques employed in construction of appliances.

Most appliances for hemophiliacs are made to plaster of Paris models. The original plaster mold should be made, whenever possible, by the orthopaedic surgeon and made so well that the positive model requires no sig-

nificant alterations. Just as the bracemaker should be present in the plaster room to see the patient and to assist in the making of the plaster of Paris mold, the orthopaedic surgeon should make frequent visits to the brace shop to observe construction of the new appliance, and supervise its fitting to the patient when it has been completed

Initial cost also militates against individualized brace construction The fact, however, that a brace is worn for a long period of time and enables a crippled person to lead a fairly normal, active and productive life, should influence any consideration of cost How very true this is for the hemophilia patient whose continuing rehabilitation at the present time depends entirely upon orthopaedic appliances. Moreover, an inadequate orthopaedic appliance will cause repeated trauma resulting in recurrent hemorrhages Here, the most expensive orthopaedic appliance is infinitely cheaper than hospitalizations, transfusions and other forms of treatment

TYPES OF ORTHOPAEDIC APPLIANCES

After individualized construction of approximately 200 major orthopaedic appliances made to plaster of Paris models, certain basic types of braces were found most suitable to the particular deformities of hemophiliacs This experience with 98 hemophilia patients has demonstrated that a certain standardization of treatment is both desirable and possible without neglecting the individual requirements of each patient

Bracing the Lower Extremities

Starting with a modernized version of the encasing molded leather-steel skeleton apparatus designed by Friedrich von Hessing in 1878, a Hessing-type leg brace has been gradually developed for the hemophilia patient

For purposes of simplification, all molded leather-stainless steel or -aluminum braces made to plaster of Paris models will be called hemophilia Hessing braces Construction details will vary depending upon individual requirements Most Hessing braces will have Lofstrand spring joints at the knee, milled ankle joints and sandal in some, knee and ankle joints are eliminated, others are made without knee joint but with milled ankle joint some have specially constructed foot sections for rigid equinus or equinovarus deformities, finally there are lower-leg, that is, below-the-knee Hessing braces with ankle spring joints for correction of equinus deformities.

In many cases, a well-padded knee cap is used in connection with the hemophilia Hessing brace, for others an elastic attachment is added for temporary introduction of small corrective forces to obtain a greater degree of extension of the knee

Occasionally a spiral-bar lower-leg brace with molded leather sandal

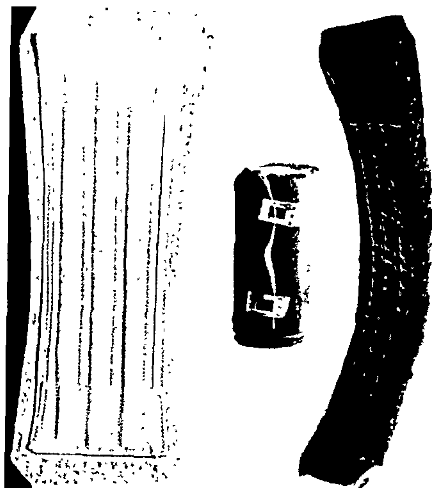


Figure 44 Posterior flexible knee splint and Ace bandage.

may be sufficient to maintain correct static alignment of the foot or to restrict dorsiflexion or plantar flexion by means of an ankle stop

Also made to plaster of Paris models are braces in which fiberglass is the chief material. These include cylinders for immobilizing the knee while the patient is bathing or swimming, and fiberglass foot plates or arch supports The latter are designed, in most instances, to correct a marked valgus position of the foot or to unweight prominent metatarsal heads and painful callosities in cases of fixed equinus deformity at a time when the major appliance has been discarded.

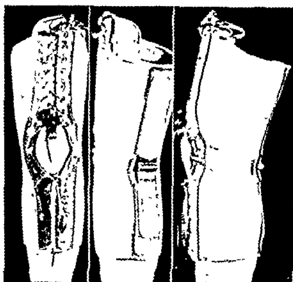


Figure 45 Hewitt-type elastic knee brace

In cases requiring unusual construction of the foot section of the brace for equinus and equinovarus deformity of highest degree, it may be impossible to obtain satisfactory fitting with a standard brace shoe A molded shoe, such as the Alan Murray Space Shoe, has been used with success when made over a plaster of Paris model of the foot section of the brace in correct static alignment of the entire extremity

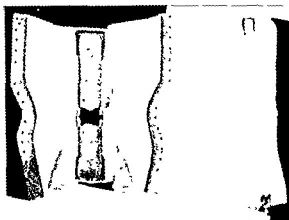


Figure 46 Hemophilia elastic knee brace with one piece foam rubber padded tongue

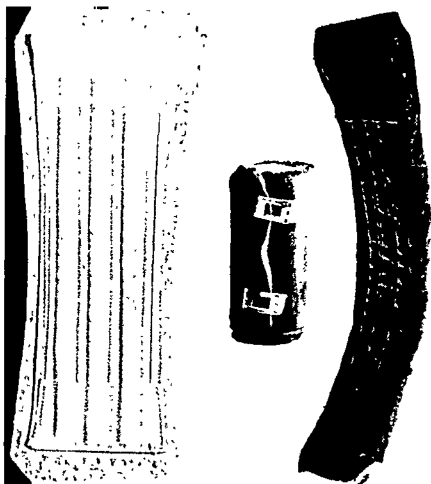


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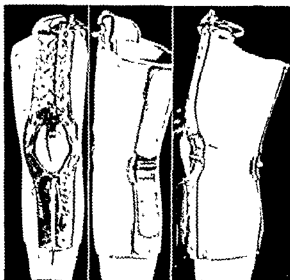


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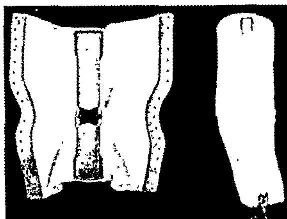


Figure 46 Hemophilia elastic knee brace with one-piece foam rubber padded tongue

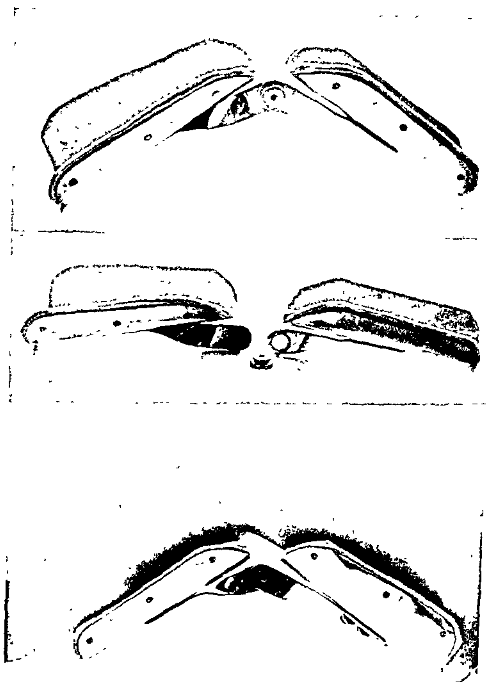


Figure 47 New Lenox Hill adjustable knee splint (hemophilia emergency splint).

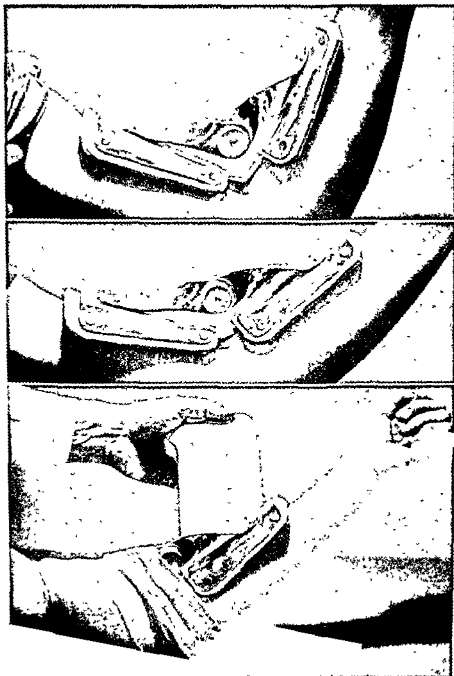


Figure 48 New hemophilia emergency splint used on the elbow

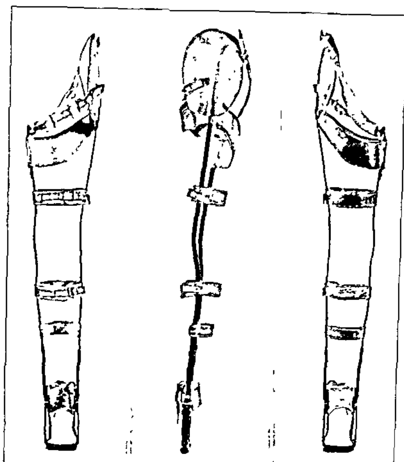


Figure 49 Jointless double-bar ischial seat leg brace with "Patten," fore-runner of hemophilia braces but not used for hemophilia.

Double-bar braces are used for unweighting the entire lower extremity by means of an ischial seat. If unweighting of the extremity is indicated, and such cases are quite rare in hemophilia, all stresses on weight-bearing should be transmitted from the region of the tuber ischii of the pelvis to the ground This means that the brace should not touch or press on any part of the extremity below the tuber ischii Inasmuch as no pressure is exerted on the leg, and the knee or ankle joints are either eliminated or locked, it is not necessary to distribute the brace over a large surface of the body. For this reason, a double-bar leg brace that is not made to a plaster of Paris model, is permissible.

The smaller but frequently used orthopaedic appliances for temporary support or for support and protection of the knee joint at night, such as

the posterior flexible knee splint and the Hewitt-type elastic knee brace, *are made to patterns and do not require a cast.*

Appliances for the Upper Extremities

As rehabilitation of the hemophilia patient calls first of all for restoration of stance and locomotion, many more appliances have been constructed for the lower extremities than for the arms.

For the upper extremities, the appliance most frequently used is the elbow brace, made to a plaster of Paris model, with fiberglass as its chief component material. This appliance has almost completely replaced the

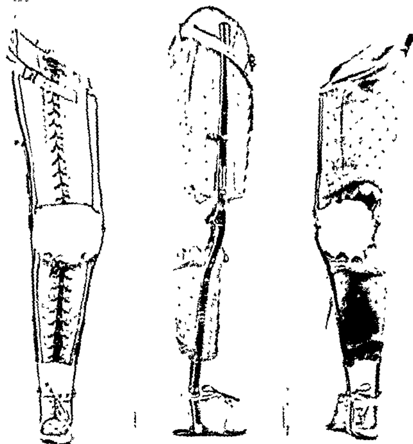


Figure 50 Hessing type long leg brace, incorporating ischial strap seat and knee lock with lever action

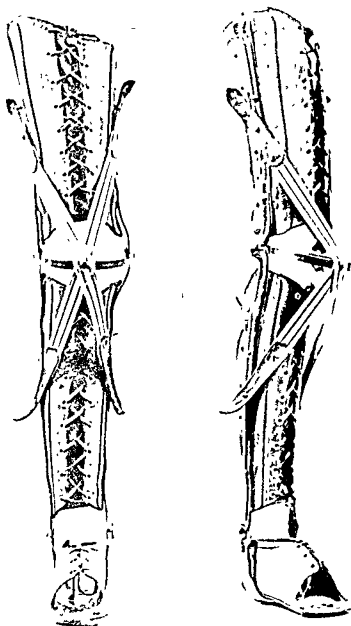


Figure 51. Hessing-type long leg brace with "artificial quadriceps"

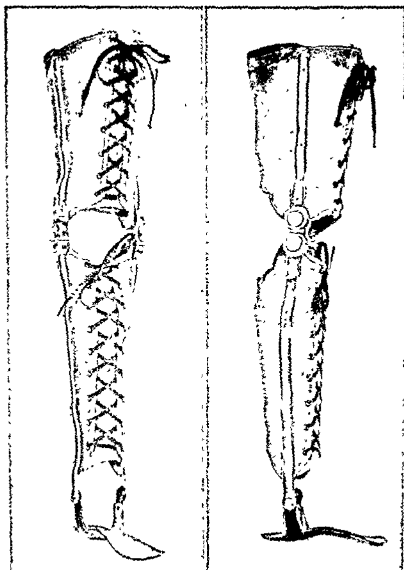


Figure 52 Hemophilia Hessing brace with Lofstrand spring joints at knee, milled ankle joints, no sandal

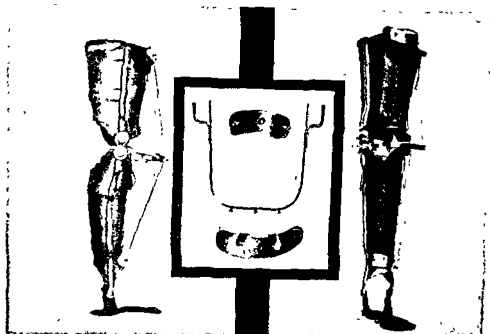


Figure 53. Hemophilia Hessing brace, same as Figure 52, with attachment for temporarily increased extension at the knee by means of elastics. Insert shows metal parts used for attachment of three #32 elastic bands.

hemophilia Hessing brace for the arm. Made for either protective or corrective purposes, fiberglass appliances are designed with or without locks, and sometimes with elastic attachments. In addition, fiberglass protective splints, without joints, may be used.

For residual contractures of forearm, wrist, hand and fingers, special splints have been highly individualized in selected cases, while Sterling Bunnell's "active splints" and simple cock-up splints for the wrist, requiring only a paper pattern, have greatly benefited many patients.

The patient must be ready for his brace before construction can proceed As a rule, this means that the greater part of a deformity must be corrected by a Quengel or wedge cast before a hemophilia Hessing brace or other major appliance may be ordered. A hemophilia Hessing brace, constructed before sufficient correction of a flexion deformity of a knee with posterior subluxation of the tibia, would be useless. Such a loss would have to be calculated in terms of both economic waste and psychological trauma to the patient who would be needlessly disappointed.

There are, however, occasional exceptions. In cases where the initial deformity at the knee joint does not exceed an angle of 150 degrees and is expected to yield rapidly to plaster of Paris correction, a model of

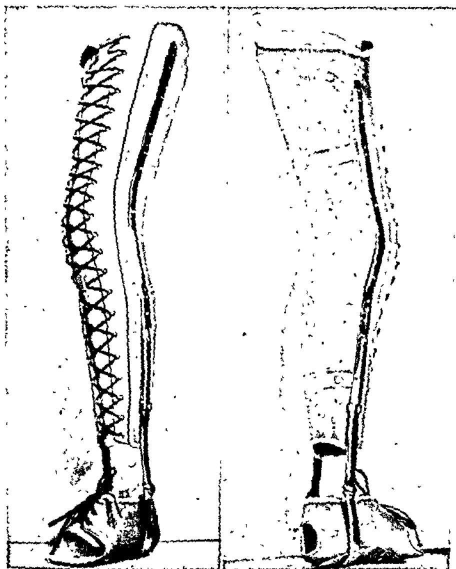


Figure 54 Hessing-type leg brace, knee joint eliminated, milled angle joint and sandal

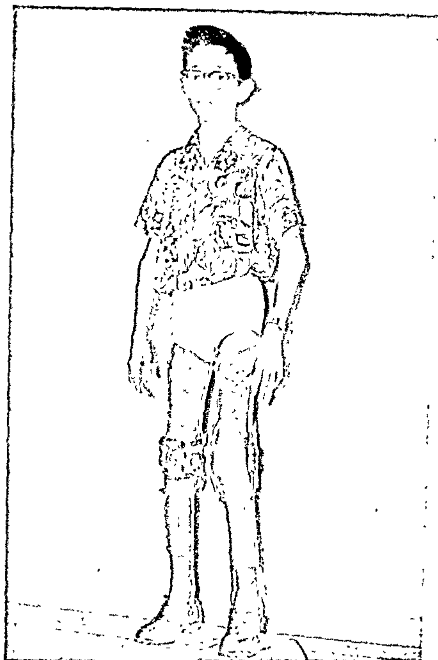


Figure 55. Two hemophilia Hessing braces (model 1951) for hemophilic arthropathies, right and left knee (*For x-rays see Figure 3*) Foam rubber padded knee cap for protection and more efficient extension of the knee is omitted on left side

Figure 56 Completely rigid equinus deformity of highest degree required special construction of foot sections of these hemophilia Hessing braces Ankle joints omitted Built-in cork wedge makes patient independent of special shoes The plantar aspect of the heel in this picture is at the level of the upper margin of shoe



Figure 57 Hemophilia lower-leg brace for correction of residual equinus deformity with Lofstrand spring joints at the ankle A separate ankle cuff is used to hold the heel to the footplate of the sandal Note the small key inserted into the Lofstrand spring joint to eliminate the spring action when the brace is put on After removal of this key, spring action forces the foot gently into more dorsiflexion

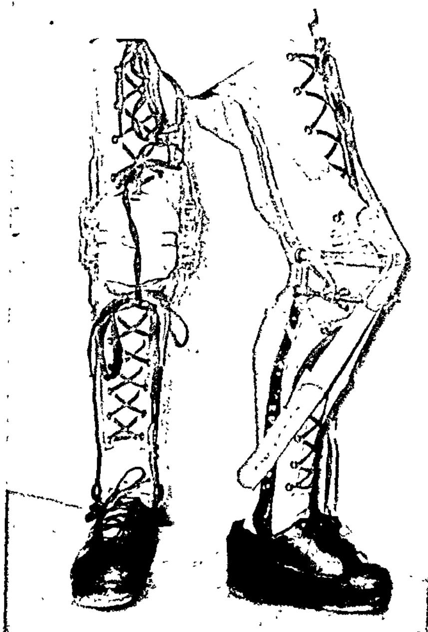


Figure 58 Two hemophilia Hessing braces on 11-year-old patient Brace for the left leg shows a new construction at the knee joint similar to subluxation hinges used in a Quengel cast for correction of posterior subluxation of the tibia This construction does not permit simultaneous use of spring joint at the knee, hence the artificial quadriceps for extension Patient wears glove molded shoes over the sandals of the Hessing braces

the leg may be made prior to application of the corrective cast, thereby saving approximately two weeks in the hospital Only rarely has it been possible to manufacture and fit a brace without initial correction of the deformity, depending entirely upon the spring action of the Lofstrand joints together with added elastic attachments to obtain the additional 20 or 25 degrees of extension at the knee necessary for good stance and locomotion

In certain instances, a force might be introduced in the hope of correcting a genu valgum or a bowing of the femur from malunited fracture while the patient grows The growing bone represents a plastic material which can adapt itself to the constant influence of a very small corrective force



Figure 59 Manufacture of a Hessing brace Left to right The plaster of Paris model has been completed The semi-circular bands for thigh and lower-leg sections have been forged to fit the model The molding leather for the sandal is already nailed to the model The molding leather is stretched over the semi-circular bands Note the development of the ischial seat in this brace and the window over the posterior aspect of the heel

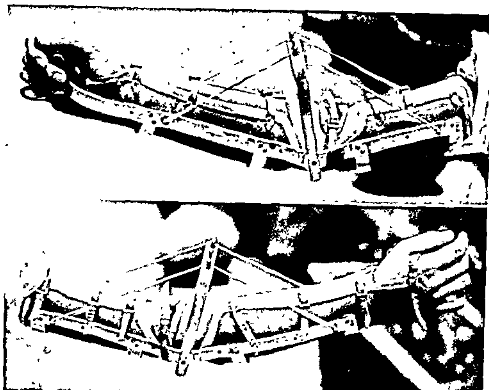


Figure 61. Arm brace for correction of extension contracture from hemophilic arthropathy at the elbow. Only two #32 rubber bands are used on each side over a U-shaped metal bridge This elastic attachment can be removed when brace is worn for protection only Note the well padded strap in the cubital region which can be moved into the most effective position as flexion at the elbow increases

In any case, an orthopaedic appliance may introduce minimal corrective forces only if it is certain that the extremity under treatment can tolerate such correction without suffering trauma Great care must be taken to avoid the use of corrective forces in the presence of ankylosis, rigid contractures or bony deformities which cannot yield to correction.

Fitting the Orthopaedic Appliance

A hemophilia Hessing brace or a fiberglass appliance cannot be finished for delivery to the patient without one or more fittings These fittings require great patience and careful handling on the part of the bracemaker, supervised by the orthopaedic surgeon. Usually a plaster of Paris maintenance cast has to be removed from the extremity to be braced, in the way described in the preceding chapter on plaster of Paris. Enough assistants

should be present to make the patient comfortable, and to hold the extremity motionless when removed from the cast and while the unlined rough sections of the new brace are applied After the fitting, the maintenance cast may be reapplied if it has been bivalved, or a new cast will have to be made. In some instances, when good correction is already present and there is little tendency toward a recurrence of flexion contracture, a strong posterior knee splint applied with elastic bandages, may suffice for the two or three working days required for completion and delivery of the orthopaedic appliance.

From what has been said about the high susceptibility of the hemophilia patient to trauma, it can be understood that the new orthopaedic appliance must have a large area of surface contact and padding at potential pressure points

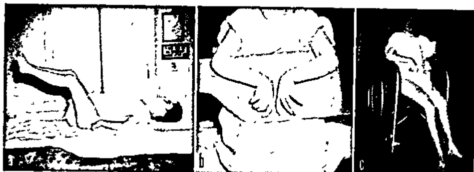
Furthermore, it is highly important that there must be no shift of the leg within the orthopaedic appliance when the position of the extremity is changed from recumbent to standing and walking. Correct alignment of brace axes with the anatomical axes of the joints in their present, frequently abnormal, position can best be checked by x-ray films of the patient wearing the brace and bearing weight on the leg under treatment These films will show at a glance if brace and anatomical axes coincide, and the exact adjustment that is necessary if they do not.

After the well-fitting orthopaedic appliance has been delivered, it is sometimes advisable to let the patient wear the brace for a day or two in bed or in the wheelchair before he starts standing and walking The orthopaedic appliance should become part of the patient's anatomy as a well-fitting denture becomes part of the oral cavity

Before standing and walking can be started, a suitable shoe must be selected to be worn with the brace At first, it is good practice for youngsters to wear a basketball sneaker before a decision can be made regarding the use of a stock surgical shoe, a brace shoe, a custom-built orthopaedic shoe, or one of the Alan Murray Space shoes made to a plaster of Paris model The basketball sneaker is quite satisfactory, even for a long period of time during the summer, fitting easily over most braces unless an unusual degree of deformity in equinus or equinovarus position is present.

In fitting a shoe, the same precautions to avoid pressure must be taken as in the manufacture and fitting of an orthopaedic appliance Usually, the shoe is in contact with the foot at the posterior aspect of the heel where pressure is not infrequent. Most hemophilia patients require softening of the counter of the shoe, and, in many instances, a stock shoe has to be fitted with a cuff to prevent the foot section of the brace from slipping In addition, crepe rubber soles are highly beneficial as shock absorbers.

At this point, the patient is ready to receive instructions for intelligent


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c


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g


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use and care of his orthopaedic appliance. Without these, perfect brace construction and fitting are useless. In formulating these instructions, the patient's age, general condition and occupation have to be considered as part of the indication

Adjuncts to Rehabilitation

There are also many factors to be evaluated in judging when canes, crutches and wheelchairs can be used as helpful adjuncts to rehabilitation. If the patient has not walked for several months or even years prior to correction of his deformity and fitting with an orthopaedic appliance, it is best to start ambulation between parallel bars or with a walker. Parallel bars are preferable because they permit better supervision of the fitting and function of the orthopaedic appliance while walking If the patient has used crutches before treatment, he may start out crutch ambulatory after he receives his appliance, discarding his crutches for one or two canes as soon as he has learned to make good use of his new brace. The use of

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Figure 62 Case #5682 (age, 15) (See Figures 25-29 for x-rays, Figure 60 for arm splint) Rehabilitation of *most severely crippled hemophiliac* Patient, age 15 (1951) to age 20 (1956). Advanced involvement of all extremity articulations from shoulder to fingers, hip to toes with bilateral bony ankylosis of ankle and foot Marked shortening of upper extremities Rigid flexion and adduction contractures at both hips Flexion contractures at both knees

(a, b and c) Condition prior to treatment (1951), (d) pelvis and right lower extremity immobilized in hip spica, left leg in plaster of Paris cast from toes to below groin First stage in correction of flexion contracture at left hip The cast was suspended in balanced traction to overhead frame of fracture bed to avoid hemorrhages into skin and subcutaneous tissues from weight of the cast Reducing the balanced traction for the cast of the left leg by one pound, a subliminal force acting constantly was created *to extend the left hip Later, the cast was wedged at the knee for correction of flexion* contracture, (e) after successful correction of flexion contracture at left hip and knee, these joints were immobilized in plaster of Paris hip spica while the right leg was prepared for a cast from toes to below groin to correct flexion adduction contracture at hip and flexion contracture at knee, (f and g) the left hip spica maintained correction of hip and knee while correction of flexion contracture at right hip was carried out similarly to that of the left in (d) Adduction contractures at both hips were then corrected with turnbuckle between the two casts (h) The right leg was fitted with hemophilia Hessing brace The left leg remained in plaster of Paris cast (i) Hemophilia Hessing *braces were fitted to both legs *Note the posterior flexible knee splint next to the right* leg which was used at night to maintain extension at the knee (j) Foot sections of hemophilia Hessing braces required no ankle joints because ankle joints were ankylosed Moderate degree of equinus deformity had to be supported by inbuilt cork wedges in the sandal to permit use of regular shoes (k and l) Patient, age 18, became ambulatory without canes wearing both hemophilia Hessing braces and fiberglass protective and corrective appliance on right arm (m) Patient obtained driver's license for specially fitted car He can handle the brake of the car with the right leg after unlocking the knee joint of the brace A special construction was needed for this purpose

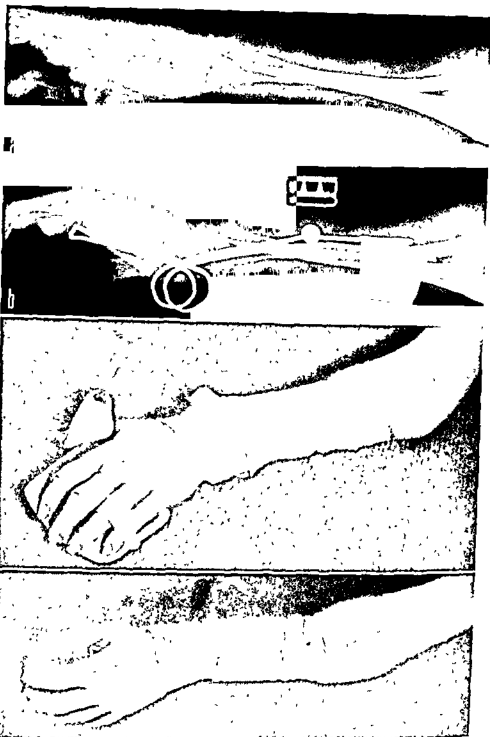


Figure 63 Case #5630 (age, 6). Volkmann's Ischemia left forearm (a and b) X-ray *control of action of Sterling Bunnell active splint for dorsiflexion at wrist* (c and d) Aluminum splints of various designs made by patient's father.

long crutches is discouraged as the trauma they inflict on the shoulder can cause temporary hemorrhages or, at worst, the development of an additional hemophilic arthropathy.

If crutches are required for a long period of time, forearm crutches are preferable to long crutches Where hemophilic arthropathies of the elbows are present, forearm crutches can be used in connection with a fiberglass protective brace with or without locking device at the elbow.

Small folding wheelchairs with adjustable leg rests and brakes are of



Figure 64 Case #5630 (age, 6) Hemophilic arthropathy right knee Hemophilic arthropathy left elbow Volkmann's Ischemia left forearm from fracture at age 18 months Treatment of hand started at age 6 (Left upper corner) Before treatment, (right upper corner and lower right and left) after treatment for one year

great value for any hemophilia sufferer no matter how good his rehabilitation. Fatigue will have to be avoided regardless of the use of orthopaedic appliances, and a small folding wheelchair is a very handy device for the patient to use periodically in house, garden or workshop even if he is capable of walking without canes. Eventually, the patient or his parents will learn to recognize the point of tolerance for physical activities without risk.

CHAPTER 5

Case Histories

WHILE INTENSIVE study of the case histories of 56 patients with hemophilic arthropathies provided the basic information for this survey, 21 of these histories have been selected to demonstrate specific aspects of hemophilic management and orthopaedic rehabilitation. Detailed and chronological presentation of these histories will reveal more dramatically than any statistical tables factors such as the value to a hemophilia patient of individually designed appliances, the possibility of orthopaedic rehabilitation despite extensive joint destruction, or perhaps the high level of orthopaedic rehabilitation after a single hospital admission

Following these 21 narrative histories, brief abstracts will appear of the remaining 35 cases comprising this study

At the back of this book, Chart I, which includes 6 pages of tabular material, records the basic statistical data from each case history

For each of these 56 patients, Chart I includes information about age at first orthopaedic examination, clinical onset of hemophilia, family history of hemophilia, occurrence of hemorrhages in cycles, internal hemorrhages, previous orthopaedic treatment, first major joint involvement and total joint involvement, degree of general disability at first examination and locomotion disability at first examination, Lenox Hill Hospital admissions, orthopaedic treatment in the hospital and on an ambulatory basis There is also data relating to rehabilitation at the end of the first active phase of treatment, general orthopaedic rehabilitation at date of last contact with the patient, and reduction in size, type or number of major orthopaedic appliances where this has occurred

NARRATIVES

Case #5112

The history of Richard O is of specific interest for three reasons First of all at the time of his initial orthopaedic consultation on April 12, 1950, treatment of hemophilic arthropathies was still in the experimental stage Chronologically the second case in our series, this was by far the more acute of the two By 1950, our general procedure for management of hemophilic joints was clearly defined, but there were still many technical details to be explored and tested His original orthopaedic appliance, for instance, was constructed empirically rather than according to established scientific principles, and he received several types of

treatment that were later abandoned in the light of more complete knowledge

Secondly, this patient's hemophilic arthropathies were very difficult to x-ray as there was extreme decalcification of all osseous structures. A-P films of the right knee could not be taken at all because of the marked flexion deformity that had developed.

Clinical onset of the disease dated from infancy with a hemorrhage into the scrotum. When the patient was 6 years old, the first articular hemorrhage involved the left knee and the flexion contracture that resulted was never fully controlled. Six years later, after severe and painful hemorrhages into the right side of the abdomen originating from the kidney, Richard was hospitalized for many weeks and developed a sensory impairment at the anterior aspect of the right leg and a flexion contracture at the knee. Bedridden or wheelchair ambulatory since that time, the boy suffered many other attacks of hematuria as well as hemophilic arthropathy of the elbows During his long and severe hemophilic history, he received no orthopaedic treatment except for knee splints at the age of 6, and only two transfusions as his veins were very poor and incisions healed slowly. Generally in better condition during summer months, the patient was able to swim twice daily and move his legs more readily under water.

Although he became chronically disabled at the age of 6, he received neither orthopaedic nor hemotological treatment during his early years.

By the time of his original orthopaedic consultation, the boy, at 14½ years of age, was negativistic, fearful, depressed, despairing of his condition and unapproachable His apprehension so interfered with initial orthopaedic treatment that three general anaesthesias for the first plaster of Paris correction of the right knee joint were needed Even after the patient had achieved an entirely different outlook on life by the time he was 20 years old, he continued to need therapy and guidance For whenever a hemorrhage threatened, his anxiety developed out of proportion to organic changes This was particularly critical as he was to suffer many internal hemorrhages, particularly into the kidneys, requiring prolonged hospital admissions and many transfusions

Although hemophilic involvement at the time of first examination also included the elbows, orthopaedic treatment was limited to the knees which were the main source of the patient's disability Clinical and roentgen examinations revealed that a moderate degree of muscle spasm increased the tendency toward flexion deformity The right knee showed an unusual degree of deformity with marked enlargement of femoral condyles and limitation of extension to 75 degrees The patella was hardly palpable between the femoral condyles The left lower extremity showed greater atrophy than the right, due to long duration of hemophilic arthropathy at the left knee. The joint was held in extension of 140 degrees.

Orthopaedic treatment was divided into several periods, the first lasting almost a year from April 12, 1950 to March 1, 1951 Prior to hospitalization the boy was treated, at the mother's request, with underwater exercises twice weekly in a Hubbard tank at the hospital to induce muscular relaxation Orthopaedic treatment was then delayed by a severe hemorrhage into the right kidney requiring

three months of bed rest. *During this time, the patient was becoming increasingly* depressed about the condition of the right leg. Thereafter, he received a series of applications of Alphatron (Radon Ointment) *to the right knee for inflammatory* changes resulting from hemorrhages. These had *no* significant effect on the condition of the joint.

The patient was finally hospitalized on November 10, 1950. The right knee, to be treated first, showed marked tenderness on palpation over the medial condyle of the femur. Active extension was not possible beyond 85 degrees. Because the patient was so apprehensive about the knee, treatment entailed a great responsibility and the psychological effect of failure upon the patient had also to be considered. Treatment was further delayed by a tonsil infection. After the patient's general condition had improved and his tension had diminished, a plaster of Paris hip spica was applied to the right leg under Pentothal sodium-gas anaesthesia. The right knee could be extended from 75 degrees to 115 degrees and at the same time, the left knee could be extended to 140 degrees. The hip spica, intended *for* wedging at the knee, was applied in two sections, one above and one below the knee joint with a cuff uniting both sections at the knee. Reaction to *the anaesthesia was so stormy and prolonged* that the left leg had to be bandaged with sheet wadding and gauze bandages to avoid injury from the cast on the right leg. *Despite these precautions, the patient suffered multiple subcutaneous* hemorrhages and bruises.

The cast, however, was *well tolerated*, and was wedged 9 *days later*. For this procedure, rectal Avertin was employed as the patient was not sufficiently cooperative to permit a wedging or change of cast without the use of a general anaesthesia. Avertin was well tolerated and the right knee could be extended to 140 degrees. At the same time, a model for a hemophilia Hessing brace with Lofstrand spring joints at the knee and simple ankle joint and sandal was made for the left leg. After removal of the wedge cast hip spica on the right leg ten days later, the knee was held in extension of 150 degrees with persistent and marked posterior subluxation of the tibia but with much less enlargement and swelling than previously noted. It was now possible to make a plaster of Paris model of the right leg to be used in the manufacture of a hemophilia Hessing brace that was jointless at the knee but included a simple ankle joint. Thereafter, a maintenance cast was applied to the right leg until appliances were delivered on January 2, 1951. The right hemophilia Hessing brace was completely successful from the start, while the brace for the left leg had to be repeatedly altered until it was finally considered satisfactory on January 11, 1951. In a very short time, the patient began to walk between parallel bars. Despite hemorrhages into right shoulder and hand, *the patient's general condition had improved considerably* while he was undergoing corrective treatment, and by the time he was discharged from the hospital on January 27, 1951 after a 78-day admission, his mental attitude had become optimistic.

A general examination on February 15, 1951 showed that the patient had gained some weight, the musculature of both thighs had increased and there was considerable improvement in the degree of calcium density at the knees

Furthermore, the boy was able to get in and out of a wheelchair without help and to perform his walking exercises with great skill and endurance. He had learned to walk amazingly well with Lofstrand forearm crutches. Support of the right leg by the jointless hemophilia Hessing brace was sufficient but the left knee continued to show a tendency toward flexion on weight-bearing. Attempting to eliminate this problem, the spring tension of the Lofstrand joints was increased As the patient was leaving for Mexico, he was also fitted with a small double-bar plastic splint to be worn at night and for swimming.

The second period of orthopaedic treatment lasted from November 5, 1951 to December 6, 1951 and included a brief hospital admission Upon examination, the patient, then 16 years old, appeared to be slightly overweight, in excellent spirits, more mature mentally and anxious to cooperate. He had suffered a few hemorrhages into the right shoulder which left no residual symptoms of hemophilic arthropathy The musculature of the upper extremities had become strong and the legs exhibited marked improvement in volume and muscle tone. Shoulders, elbows, knees, feet and ankles showed most satisfactory recalcification of osseous structures. There was some decalcification throughout the head of the right humerus but no evidence of involvement with hemophilic arthropathy at shoulder joints. During a 5-day hospitalization, plaster of Paris models were made for a new light-weight hemophilia Hessing brace with Lofstrand spring joints for the right leg, and a hemophilia Hessing brace similar to the original one, but also lighter in weight, for the left. These appliances, delivered on November 29, were immediately satisfactory and the patient was well equipped to leave for South America where his family planned to live for approximately 2 years.

On March 31, 1954, when Richard returned for his third period of treatment, he had grown considerably and was extremely mature for his 18 years. Though mostly cane or wheelchair ambulatory, he was able to stand without appliances. He had had no significant trouble from hemophilic arthropathies in the interim, with only an occasional swelling of the right or left elbow after using his canes too much He had suffered, however, a severe cut of the left fifth finger, losing [illegible] on at the [illegible] leveloped [illegible] ted with intermittent kidney hemorrhages

The right leg was remarkably stable and the knee had remained asymptomatic and extended to 165 degrees The left knee showed AGE 140 degrees. There was some restriction of motion at the ankles with dorsiflexion limited to 100 degrees on the right and 90 degrees on the left. Occasional episodes of swelling and pain at the right ankle from a small hematoma had occurred because orthopaedic appliances had begun to wear out and required replacement. Otherwise, they had served their purpose very well Even climbing 26 steps in order to attend high school had caused no discomfort Plaster of Paris models were subsequently made for new hemophilia Hessing braces with Lofstrand spring joints at the knee Extension of the right knee was limited to 165 degrees, of the left

knee to 145 degrees The appliance for the left leg was also fitted with an elastic attachment for increased extension at the knee Delivery of the new appliances, delayed by an intercurrent hematuria and a hemorrhage into the right shoulder, was finally made and the patient wore them comfortably with new shoes with shock-absorbing crepe rubber soles Treatment during this period also included aspiration of the blister on the left fifth finger

At an interval examination on August 24, 1954, the patient stood and walked better with his braces than ever before He had suffered no further hemorrhages internally or otherwise since April 1953 Correction of the eversion of the right foot, intended by the new brace, was fully accomplished The left knee remained flexible to AGF 90 degrees while the right was extended to 170 degrees The musculature of both legs was greatly improved. Rehabilitation had proceeded to the point where the patient, about to enter college, had obtained his driver's license, swam regularly, and was able to take a few steps without braces

When next examined on April 4, 1955, the boy's general condition was excellent and he had experienced no hemorrhages or other disabilities while in college The left knee joint and the left quadriceps functioned so well that the springs of the Lofstrand knee joints were removed

Ten days after this satisfactory examination, Richard was hospitalized with an acute hemorrhage into the left groin or the left lower abdominal quadrant with marked secondary anemia, and complaints of severe pain in the left hip region The impending flexion contracture was almost completely prevented, however, by proper positioning in bed. By April 23, 1955, after a 9-day hospital admission, the patient had recovered sufficiently to leave the hospital crutch ambulatory with only a slight residual flexion contracture at the left hip His secondary anemia was entirely corrected This brief episode of bleeding into the soft tissues of the left hip and lower abdominal region reversed rehabilitation of the left leg as it was followed by weakness of the left quadriceps and by sensory impairment on the dorsum of the left thigh It was therefore necessary to reapply the Lofstrand springs to the hemophilia Hessing brace in highest tension On May 1, 1955 the patient was able to return to college crutch ambulatory and permitted to lead a sedentary life

When seen on August 24, 1955, he appeared to have made a good recovery, walking and driving once again, although he continued to complain of hypesthesia and paresthesia on the dorsum of the left thigh and of persistent weakness of the left quadriceps Motion of the left knee joint ranged from AGE 150 degrees to AGF 70 degrees With the Lofstrand spring joints in highest tension, he had enough strength to handle the brake of his specially equipped car without risk He had had no significant hemorrhages although he suffered a hematoma at the right ankle and a short episode of hematuria

On November 25, 1955, an excellent report was received from the patient's mother Although the springs of the Lofstrand knee joints on both braces had been broken, ambulation was much improved without them

Seen on April 3, 1956 during his Easter vacation, the patient's general condition was better than ever and he was in very good spirits His posture had

improved and he had been walking as much as three city blocks without pain and undue fatigue. As both hemophilia Hessing braces were in poor state of repair and required immediate replacement, plaster of Paris models were made of both legs for new appliances

Hardly more than 24 hours after this examination, the boy suffered an acute locking of the left knee with considerable pain when getting out of a taxicab The joint was held in flexion of 140 degrees Relapsing into a state of complete anxiety, he refused to move the left knee because he was afraid of a hemorrhage. Sedation and an attempt to relax the left leg by prolonged hot baths failed At this time the patient's mother reported that he had been undergoing psychotherapy with some success but that he seemed extremely apprehensive whenever he came on a visit to New York City When examined on April 5, he was almost hysterical, refusing to move his left knee He would not wear the brace and was unwilling to get out of his wheelchair. Because of his neurotic reaction to the condition of the left knee, he was promptly admitted to the hospital.

Roentgen examination of the left knee joint showed no soft tissue swelling or evidence of fluid Comparison with films taken in 1951 showed some improvement in the appearance of the osseous structures with increased calcium density and better trabeculation The articular surfaces, although bare of cartilage, appeared more smooth

Under complete bed rest, sedation, injections of Demerol® and psychotherapy by his psychiatrist, the patient's general condition as well as local findings at the left knee gradually improved until he was able to reapply the hemophilia Hessing brace By April 11, 1956, he had lost most of his fear and anxiety. Extension of the knee was again possible to 150 degrees and he was discharged from the hospital after a 6-day admission Returning to college the next day, he remained in the infirmary for a few days until he had regained his confidence and was able to walk as usual. On May 11, 1956, the new appliances for the lower extremities were fitted and delivered.

The latest report indicated that the boy was getting along very well with his new appliances. He was even able to change a tire on his car without any assistance and without undue aftereffects

From time to time the psychological aspect of this case has been carefully reviewed Although this is not the province of the orthopaedic surgeon, its relationship to orthopaedic treatment of hemophiliacs must be considered It is significant that a remarkable change in the mental attitudes of this patient occurred after the first period of rehabilitory treatment, particularly during his stay in Mexico, where he was able to lead a more active life. He became quite optimistic although periods of depression with marked anxiety were still not infrequent at the first signs of hemorrhage He made great progress during his 2-year stay in South America where he engaged in a social life with parties, swimming and games At 18 years of age, he was much more mature and could take his fate with more equanimity. But even at the age of 20, after 2 successful years in college, he still required psychiatric treatment to overcome anxieties such as were in evidence during his last hospital admission in April 1956

Case #5637 (See Figure 39)

At the time of first orthopaedic examination on August 1, 1951, Alan S, age 11, had been wheelchair or crutch ambulatory for 5 years because of a severe flexion contracture of the right knee The patient was one of four children and his only brother had died after a tonsillectomy at the age of 7 years There were no known hemophiliacs in the family's ancestry

Apart from abnormal bleeding after circumcision and at 9 months of age when he received his first transfusion, he suffered no serious hemorrhage until he was 6½ years old At that time, a definite injury to the right knee led to hemophilic involvement of the joint which became markedly swollen, painful and filled with blood During the next 6 months, the boy developed a flexion contracture of the knee which was treated by manipulation under anaesthesia, followed by application of a plaster of Paris cast which remained in place for six months While the leg was immobilized, the patient complained of severe pain caused by pressure from the cast at the heel and the instep of the right foot When the cast was finally removed, a decubitus was noted at the heel but the flexion contracture of the knee had not improved No further treatment was offered and the patient remained wheelchair ambulatory for eighteen months Thereafter, he became crutch ambulatory although he had not borne weight on the right leg since the original injury During his childhood, he also suffered hemophilic involvement of the elbows as well as an acute episode of gum bleeding His parents claimed that he tended to bleed more readily toward the end of March or the beginning of April

Clinical examination of the right leg revealed considerable atrophy of the musculature in comparison with the left side. The knee joint showed a range of motion from AGE 125 degrees to AGF 75 degrees Roentgen examination of the knee showed a severe hemophilic arthropathy with deformity, considerable valgus position and marked posterior subluxation of the tibia in relation to the long axis of the femur. Articular surfaces of both femoral and tibial condyles were irregular with more thinning of the articular cartilage throughout the lateral compartment of the joint. Femoral condyles showed more deformity and irregular trabeculation with sclerosis of the bone close to the epiphyseal line than to the tibial condyles There was generalized decalcification of the skeleton and underdevelopment of the tibia Films of the left knee joint revealed impairment of growth of the tibia but no evidence of hemophilic involvement

Elbows exhibited a tendency toward cubitus valgus with restriction of extension to 165 degrees on the left side and 175 degrees on the right

During a 59-day hospital admission beginning August 8, 1951, a plaster of Paris hip spica was applied to the right hip and leg under Avertin anaesthesia and prepared for wedging at the knee Although the cast was not tolerated and the knee section had to be removed, the hip spica was utilized for preliminary correction of the marked posterior subluxation of the tibia by means of balanced traction in a fracture bed with overhead frame. It was important that some correction of this deformity be obtained prior to treatment of the flexion contracture

of the knee in order to avoid additional damage to the articular surface of the femoral condyles Furthermore, this procedure served to decelerate the pace of orthopaedic treatment as the patient's psychological difficulties had increased and he seemed to require more time to adjust to the hospital environment and to treatment.

After the wedge cast hip spica was removed to below the knee, a single lateral x-ray film showed decreased posterior subluxation of the tibia. A Quengel cast hip spica, with subluxation hinges at the knee, was then applied and correction started with the knee held in extension of 135 degrees. The cast was well tolerated but progress was extremely slow because of the unusual degree of posterior subluxation of the tibia and the close contact of the deformed articular surfaces of femur and tibia as well as extensive soft tissue contractures The special subluxation joints, connecting thigh and lower-leg sections of the cast, permitted simultaneous extension of the knee joint by regular Quengel, and forward motion of the tibia in relation to the femoral condyles.

After extension of 150 degrees at the knee had been obtained, the Quengel cast was removed in sections to prevent strain on the knee, and the joint was immobilized first with a dorsal splint and then in a maintenance cast As clinical and x-ray studies had indicated that it would be impossible to proceed with further plaster of Paris correction without a period of rest, the patient was discharged crutch ambulatory on October 6, 1951 with his right knee extended to 150 degrees and the greater part of the posterior subluxation of the tibia corrected He was advised that he might soon return to school.

On November 16, 1951, the maintenance cast on the right leg was removed and a second period of orthopaedic treatment, this time on an ambulatory basis, was begun with application of a new Quengel cast with subluxation hinges at the knee. In short order, correction of the posterior subluxation of the tibia had proceeded as far as the appliance would permit, although at the same time there had been some loss of extension at the knee Three brass hooks were then incorporated into the proximal and distal sections of the cast with three No 32 elastics fastened to the hooks to serve as uninterrupted subliminal forces for further extension. A bridge at the knee was also inserted to improve the efficacy of the corrective forces. The boy, however, had applied too many elastic bands in an effort to straighten the knee as much as possible. The cast had caused too much pressure on the knee and cut sharply into the skin below the gluteal fold Although this had occurred during a period when the patient exhibited very little tendency toward hemorrhaging, and there was no bleeding at all from the laceration, a plasma transfusion was administered as a matter of prophylaxis, and the cast was trimmed.

On December 10, 1951, it was finally possible to take the plaster of Paris model for the patient's first hemophilia Hessing brace, holding the right knee extended to 165 degrees. The appliance, constructed with Lofstrand spring joints at the knee, simple ankle joints and sandal, was delivered on December 27 after the necessary padding of pressure points with foam rubber The patient was immediately able to walk, using one cane, and to return to school

Examined several times in the early part of 1952, the patient appeared in excellent general condition despite a hemorrhage into the left calf, and he had no pain or other symptoms at the right knee The area of decubitus below the right gluteal fold was well healed and he was completely ambulatory without a cane, holding the right knee at AGE 155 degrees

On March 28, the boy complained of an increasingly severe headache and shortly thereafter had convulsions and became unconscious. He was subsequently taken to the hospital by ambulance in critical condition with an intracranial hemorrhage During a 2-week admission, he was subjected to necessary precautionary treatment that included oxygen, gastric lavage, transfusions with quick-frozen plasma, and Dilantin®, and made a complete recovery Neurological examination and an electro-encephalogram showed that the hemorrhage was probably extradural with a possible affection of the left occipital lobe Throughout this enforced rest, the patient continued to wear the hemophilia Hessing brace with added elastics, with beneficial results.

The patient had few hemophilic problems in the months preceding his next admission to the hospital for orthopaedic treatment At the end of May 1952, he was hospitalized for 3 days for slight gum bleeding which quickly subsided after a transfusion and refrigeration

At an examination in August 1952, the right knee demonstrated the return of a considerable degree of flexion with a range of motion from AGE 160 degrees to AGF 130 degrees The musculature of the right leg, particularly the calf, had so increased in volume that construction of a second hemophilia Hessing brace was required

On March 23, 1953, the boy was admitted to the hospital for further correction of the flexion and valgus deformities of the right knee and residual posterior subluxation of the tibia Although the knee had remained asymptomatic for a year, there had been no additional improvement in the degree of extension available from the Lofstrand spring joints The valgus deformity was still pronounced Roentgen examination showed sufficient articular space to allow further correction of the flexion deformity without causing impact of the tibial condyles against the femur

During a 14-day admission, the residual flexion deformity of the knee yielded surprisingly quickly to the corrective force of a large Quengel cast with subluxation hinges at the knee When the cast was removed on April 7, 1953, the joint was completely extended although residual posterior subluxation of the tibia detracted from complete extension At this point, correction of the valgus deformity could not be accomplished without surgery The old hemophilia Hessing brace was readjusted, holding the knee in complete extension, and the patient was discharged from the hospital completely ambulatory

When next examined on July 14, 1953, the boy had grown considerably and had lost weight at the same time His general condition remained excellent and his gait had continued to improve It was then decided to utilize this period of rapid growth for correction of the marked genu valgum deformity The hemophilia Hessing brace was accordingly fitted with a special leather knee

cap to be attached to the external Lofstrand knee joint. It was planned that gradual tightening of the cap would correct the valgus position of the knee Three months later, the patient had grown so much that a larger hemophilia Hessing brace was required As the knee cap had been quite successful in correcting part of the valgus deformity, it was to be worn with the new appliance

At a further examination on February 27, 1954, the appearance of the right leg was better than ever The new orthopaedic appliance had been successful in completing extension to AGE 180 degrees despite persistent and marked posterior subluxation of the tibia, and the valgus deformity of the knee had been gradually decreased

In August 1954, the patient was advised to discard the hemophilia Hessing brace at night The leather knee cap, however, was to be worn with the brace as the small force introduced by this attachment could still be utilized to decrease the valgus deformity as long as growth continued.

On January 15, 1955, the brace was reconstructed to further eliminate the tendency toward genu valgum deformity The knee cap was discarded as it had started to interfere with flexion of the knee, and a pressure pad was added to the hemophilia Hessing brace above and below the medial condyle of femur and tibia. The patient reported at that time that he was attending a preparatory school, and travelling comfortably back and forth by bus

By March 17, 1955, the condition of the knee was further improved with a range of motion from AGE 180 degrees to AGF 140 degrees. Because the musculature of the right thigh had increased in strength, spring tension in the Lofstrand knee joints was removed

Aside from an 8-day hospital admission in the fall of 1955 for general debilitation and for pain in the right hip, there were no physical complications throughout the year The patient received a new hemophilia Hessing brace in December

When last observed on June 19, 1956, he had grown considerably but his hemophilia Hessing brace had remained comfortable. Orthopaedic rehabilitation was considered complete as the right knee joint had been free of pain and other hemophilic symptoms since the first hemophilia Hessing brace had been applied 5 years earlier In addition, the valgus deformity had finally yielded to treatment and posterior subluxation of the tibia had been almost eliminated Exhibiting remarkable physical endurance, the patient was able to stand and walk with great facility as the function of the right knee joint had been largely restored

Case #5630 (See Figures 16, 63 and 64)

When James C, age 6, appeared for his first orthopaedic examination on August 3, 1951, he had been totally disabled for 10 days following a hemorrhage into the right knee caused by an injury to the joint. Examination of the knee revealed moderate swelling, evidence of free fluid, and a range of motion from AGE 140 degrees to AGF 60 degrees

As there had been no family history of hemophilia, extensive bleeding at circumcision revealed the existence of the disease Other hemophilia symptoms

had been mild until the right knee was affected when the child was 5 years old. Generally, the boy's bleeding tendency seemed more pronounced at the beginning of summer.

The remarkable feature of this case history, however, was not the hemophilic arthropathy of the right knee joint but rather the Volkmann's contracture of the left forearm which had developed after the patient, at 18 months of age, had caught his left forearm, wrist and hand in the wringer of a washing machine. After more than a year of unsuccessful physical therapy, the parents of the child were told that the left arm was a complete loss and no longer susceptible to treatment. This diagnosis was fully accepted and the condition of the forearm was henceforth de-emphasized.

Examination of the left upper extremity showed a typical Volkmann's contracture with maximum flexion deformity of wrist, hand and fingers Although the hand was useless, there was no evidence of neurological involvement The elbow was slightly swollen with no significant restriction of motion and the shoulder was freely movable

Roentgen examination of the right knee showed moderate generalized decalcification of osseous structures Soft tissues showed increased shadow density and fusiform enlargement The patella was also markedly enlarged. Articular surfaces of femoral and tibial condyles showed moderate roughening with small areas of bone destruction or compression at the medial condyle of femur and tibia Epiphyseal lines were well preserved, and there was no evidence of bone involvement beyond the epiphyses Osseous structures of the left knee were almost normal with the exception of a few areas of decreased calcium density in the lateral condyle of the femur

X-ray films of the left elbow demonstrated a striking example of cystic bone destruction caused by hemorrhages There was considerable thinning of cartilage with irregularities of articular surfaces of ulna, radius and the lateral condyle of the humerus. The skeleton of the left forearm showed atrophy and a shortening in comparison with the right side Large cystic areas were noted in the proximal end of the ulna involving both coronoid and olecranon There was moderate decalcification of all osseous structures of the left hand and wrist with the joint held in flexion of 85 degrees as a result of contracture There was no evidence of hemophilic arthropathy distal to the elbow

The patient was hospitalized on August 8, 1951 for treatment of the right knee and left forearm His parents were informed that carefully planned orthopaedic treatment, incorporating many types of corrective techniques such as Quengel and wedge casts, active corrective splints and fiberglass braces, with periodic immobilization of the extremity in maintenance casts, would lead to considerable recovery of the function of the left hand Fortunately, the patient's father, a highly skilled technician, was competent to build appliances for his son and maintain them in good working order

The flexion contracture at the right knee presented no problems After a plaster of Paris model was made of the leg with knee held in maximum extension of 160 degrees, the contracture was completely corrected by a wedge cast

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The flexion contracture at the right knee presented no problems After a plaster of Paris model was made of the leg with knee held in maximum extension of 160 degrees, the contracture was completely corrected by a wedge cast

As soon as the patient was fitted with a hemophilia Hessing brace with Lofstrand spring joints at the knee, he was able to walk with his knee fully extended, and without the aid of crutches or canes.

Treatment for the deformity of the left forearm began with application of a Quengel cast from elbow to metacarpophalangeal joints The wrist yielded more rapidly than anticipated with only 45 degrees missing to complete extension at the time of the patient's discharge on September 8, 1951 after a 31-day admission

For the next 6 months, the cast was worn continuously. By mid-November, the contracture at the wrist, and deformities at metacarpophalangeal and proximal phalangeal articulations, had been completely corrected X-ray films taken February 7, 1952 showed that extension of wrists, hands and fingers had been completely maintained and that there was better calcification of osseous structures

A setback occurred in March 1952 when a hemorrhage into the left elbow required immobilization of the joint in a well-padded plaster of Paris cast At the same time, the patient was instructed in exercises to be performed several times daily using a knuckle bender splint A rubber bandage was also used each day to force fingers into complete flexion. After the cast was removed, splinting of the forearm was followed by the use of a Sterling Bunnell active cock-up splint for the left wrist until the musculature of the arm was tested and showed some response to electrical stimulation At this point, the patient was given a portable electric machine with small electrodes to be used at home for daily stimulation of extensor and flexor muscles of the forearm. (This procedure was followed successfully until March 1953)

During this period, the right knee had remained fully extended and asymptomatic and a new hemophilia Hessing brace was manufactured and delivered

On February 24, 1953 a new series of treatments was started for the left forearm, wrist and hand using plaster of Paris casts and orthopaedic appliances for correction of the residual deformity and contracture due to the shortening of flexor tendons Elastic traction, applied to the fingers by means of small leather loops, yielded surprisingly good results by March 7, 1953 and two weeks later the four long fingers of the left hand were completely extended Thereafter, only night splinting and exercises for flexion were necessary In September and October of the same year, further treatment included the use of a two-piece fiberglass brace with universal joint at the wrist, to improve dorsiflexion and radial abduction

By March 13, 1954, roentgen examination of the left elbow and forearm showed increased extension at the elbow to 165 degrees, no further flexion deformity and only slight ulnar deviation of the hand due to shortening of the ulna All bony structures appeared better developed and calcified

Additional x-ray films taken on December 4, 1954 showed tremendous improvement in the appearance of osseous structures of the left elbow with a marked degree of recalcification and retrabeculation. Large juxta-articular cysts were mostly filled in with bone to the extent that they were hardly visualized, and bones were better calcified. Although there was still considerable decalcification of the left wrist and hand, all fingers were well extended

Films of the right ankle showed skeletal underdevelopment in comparison with the left side Also, the valgus position of the astragalus within the mortise, which existed at both ankles, was slightly more prominent on the right side.

Despite considerable improvement at the left elbow, additional rehabilitation of the forearm was desirable and a fiberglass brace was constructed for the left wrist and hand to provide increased flexion of fingers until the fist was complete Roentgen examination had also indicated preservation of structures of the right knee, ankle and foot but hemophilic arthropathy was sufficiently marked there to warrant continued support although the patient had walked a good deal without his orthopaedic appliance during the summer A plaster of Paris model was therefore made of the right leg for a new hemophilia Hessing brace

Following a series of hemorrhages into the right elbow, the joint was examined on November 17, 1955 and found to be considerably enlarged There was crepitation on active and passive movements and a range of motion from AGE 170 degrees to AGF 90 degrees X-ray films showed quite marked pathologic changes with considerable decalcification, small juxta-articular cysts, and some increased soft tissue around the joint

In January 1956, the patient received an aluminum brace for protection of the right elbow together with another hemophilia Hessing brace for the right leg and, for the left arm, a fiberglass appliance with a joint at the elbow and a universal joint connecting the forearm section with the hand section for further correction of the wrist Three months later, a fiberglass appliance similar to that worn on the left side was made for the right elbow

Throughout a five-year period of observation, treatment of this patient was complicated by disabling hemorrhages into the left thigh, left calf and the elbows The patient had also suffered a serious hemorrhage in September 1952 into the left iliopsoas muscle which required prolonged bed rest and transfusions with whole blood and plasma Development of a flexion contracture at the left hip had required positional treatment in bed with a sandbag on the left buttock while the patient was prone until extension at the hip was possible to 165 degrees and there was no more pain

The left ankle had also been affected and roentgen examination in October 1954 had revealed a moderate degree of generalized decalcification of the joint and slight osseous changes suggestive of an early stage of hemophilic arthropathy

Despite these difficulties, treatment proceeded according to a well defined plan While correction of the crippling flexion contracture of the right knee by a wedge cast, and maintenance of permanent rehabilitation in a series of hemophilia Hessing braces were fairly routine, the rewarding feature of the case was rehabilitation of the maimed left arm for which all hope had previously been abandoned

As the family had to move from New York City, recent contact with the patient is not as frequent as it had been during the first 3 years of treatment Although the boy's condition has remained satisfactory, even greater rehabilitation of the function of the left forearm might be possible if examinations could be made at more regular intervals

Case #5882 (*See Figures 25, 26, 27, 28, 29, 60, 62*)

An outstanding illustration of rehabilitation in this series, Howard S. was the only patient under treatment with hemophilic involvement of every articulation of the upper and lower extremities When first examined on October 4, 1951 at age 15½, he was totally disabled and required transportation by ambulance Crippled by hemophilic arthropathies, he could not even tolerate wheelchair ambulation as he was unable to maintain his balance in a sitting position

With a family history that was negative for hemophilia, he suffered his first hemorrhage at circumcision. During infancy, he sustained face injuries with multiple cuts and lacerations when he fell through the windshield of a car, and again when he crashed into a mirror while climbing on his mother's dresser.

Hospital admissions during childhood and early adolescence were mostly for articular involvement although he also had many internal hemorrhages, with bleeding from the urinary tract being the most difficult to control. Numerous attempts to treat his deformities by manipulation under anaesthesia and with plaster of Paris failed.

Initial clinical and roentgen examinations revealed advanced changes in all articulations of the upper and lower extremities with varying degrees of contractures due to involvement with hemophilic arthropathy. All osseous structures showed marked decalcification and lack of trabeculation Knee joints showed 90 degree flexion contractures and hips showed flexion and adduction contractures. The presence of symmetrical bony ankylosis of feet and ankles in equinus position was not related to hemophilic arthropathies but was caused, in all probability, by rheumatoid arthritis.

Shoulders showed restriction of motion and both arms were conspicuously short with elbows in flexion and forearms showing pronation contractures Wrists were stiff, and all fingers were deformed.

Treatment following these examinations included only two hospital admissions specifically for plaster of Paris correction That extensive orthopaedic treatment could be carried out largely on an ambulatory basis was due to the fact that the ambulance of the fire department in the community where the family lived was made available to the boy without expense whenever changes of casts were required at the hospital

During the first admission from November 6 to November 27, 1951, a Quengel cast hip spica was applied under Avertin anaesthesia to the left lower extremity with the hip, knee and foot held in maximum flexion to immobilize the pelvis for correction of the flexion contracture at the hip. After some degree of correction was obtained, a similar cast was applied to the right side for the same purpose

Because of the unusually complicated nature of this case, simultaneous correction of deformities at hip and knee on the side under treatment necessitated the use of such large and heavy plaster of Paris casts that the weight of these casts alone could have caused pressure sores, bruises and hematomas in soft tissues. Such injuries were avoided by placing the patient with his hip spica in a

fracture bed and suspending the entire cast in balanced traction It was possible to use gravity instead of a Quengel for gradual correction of the flexion deformity at the hip by reducing the balanced traction for the leg under treatment by just one pound. Adduction contractures were eventually corrected by placing turnbuckle screws between the casts on both thighs

Treatment of hips and knees was further implemented by a succession of wedge cast hip spicas Plaster of Paris models were then taken of both legs for hemophilia Hessing braces and wedge casts were again applied until the appliance for the right leg could be fitted. After a 21-day admission, the patient was discharged from the hospital with the new appliance for the right leg and a wedge cast for the left

Examined 4 days later, extension at the right knee had reached 170 degrees with some valgus, and hips had maintained some degree of abduction and extension After fitting and delivery of the left hemophilia Hessing brace, extension to 150 degrees was quickly obtained at the knee but there was a marked tendency toward flexion under weight-bearing

Within two months, extension at both knees had reached 170 degrees and the patient was capable of sitting at a desk By July 25, 1952, he had begun to stand with crutches As no attempt could be made to correct the feet and ankles in the equinus position in which they were ankylosed, special shoes were designed with compensatory cork wedges to serve as a platform for the regular foot sections of the hemophilia Hessing braces

A second hospital admission, required during this period for oral surgery under general Avertin anesthesia, was uneventful

Although the patient remained free of hemorrhages for the next three months, attempts at crutch ambulation were unsatisfactory as the left shoulder developed great pain and the patient was bedridden for extended periods after using crutches for only a short time While the left leg maintained a satisfactory degree of extension and supported the body weight, the right leg was held in poor static alignment due partly to restriction of extension at the knee to 155 degrees and partly to the fact that the right shoe did not have the correct wedge A walker was therefore temporarily substituted for crutches, and elastic attachments for increased extension of the right knee were provided for daily use By January 21, 1953, the patient was finally able to stand quite well with two crutches As the experimental shoes had not been successful in compensating for the equinus deformity of feet and ankles or in providing stability, it was decided to build new lower-leg sections for the hempohilia Hessing braces incorporating fiberglass foot plates and cork wedges to compensate for the permanent equinus position Once the entire compensation became part of the orthopaedic appliance, it was easy to fit the patient with regular oxfords The reconstructed appliances afforded better stability, and the patient, ambulating comfortably with crutches, was free of pain at the left shoulder, despite x-ray evidence of advanced destruction of the humeral head By July 9, 1953, the patient exhibited remarkable progress, walking long distances without outside support of any kind and beginning to participate in physical activities such as

miniature golf. Rehabilitation to this point had been more rapid and complete than anticipated

On July 19, a second hospitalization was begun for further correction of the right knee which had retrogressed to AGE 150 degrees, as well as for initial correction of the flexion contractures of the right elbow, pronation and supination contractures of the right forearm and marked deformity of the right wrist and hand

Roentgen examination during this 10-day hospital admission revealed definite recalcification of the left shoulder since crutches had been discarded.

After a plaster of Paris model was made of the right arm to be used in the manufacture of a corrective orthopaedic appliance, a wedge cast was applied to the entire right leg and was successful in obtaining extension at the knee to 165 degrees This correction, combined with delivery of a pair of reconstructed hemophilia Hessing braces, provided considerable improvement in the patient's gait and he was discharged as soon as he received a fiberglass appliance for his right arm

During the fall and early winter of 1953, the patient was hospitalized for a long period of time and required many transfusions for intestinal hemorrhages He had recovered sufficiently by the beginning of 1954 to continue with school and to work at a photography store

When next examined on September 2, 1954, the patient reported that he had taken a few steps without his appliances Comparison of new x-ray films of the entire skeleton with those taken on July 20, 1953 revealed considerable improvement in calcium density and in clarity of structural details, attributable to the patient's increased activities The left shoulder showed considerable healing While the left knee was extended to 160 degrees, the right showed no significant improvement in extension although the patient was able to walk quickly and with great stability It was then decided that the springs of both knees were to remain on high tension and the right knee was to be further corrected by daily use of three rubber bands worn over a knee bridge for at least one hour The fiberglass brace for the right arm, which had been incorrectly used and then discarded as it had interfered with circulation in the hand, was to be worn at night using only one rubber band for correction of the flexion contracture at the elbow.

Periodic examinations during 1955 indicated progressive rehabilitation of stance and locomotion Leading a very active life, the patient was able to take full charge of the photography store where he was employed, and had received his driver's license Since he had started to drive, range of motion at the knees had noticeably increased By the end of the year, examination of the lower extremities showed extension at the knees to 165 degrees with a moderate degree of posterior subluxation of the tibia in relation to the femur and some degree of valgus and external rotation of the lower legs At the time, the patient considered manipulation of stairs to be his main difficulty. Plaster of Paris models were taken of both legs for new hemophilia Hessing braces and for better fitting of the molded leather sandals, separate models were taken of the feet. Upon delivery, braces were locked at the knees in maximum extension in

an effort to provide additional stability. However, as the patient was unable to drive with the right knee extended, the right hemophilia Hessing brace was fitted with a special device allowing the patient to unlock the knee joint of the brace through his trousers whenever he wished to drive.

When last examined in May 1956, the patient, at 20 years of age, exhibited rehabilitation surpassing that of many patients with lesser hemophilic involvement Almost independent, physically as well as financially, he managed to get around very well and his arms had become longer and more useful Working successfully as a press photographer, he was able to walk many miles to fulfill an assignment and to drive his specially equipped car with great endurance over long distances. He was also about to marry

Case #5733 (See Figure 10)

Although there was no hemophilia in John P's ancestry, his older brother was a hemophiliac and had died of the disease The patient was therefore not circumcised, and did not experience any unusual bleeding until his upper lip was injured at 4 years of age. Since then, he required frequent hospitalizations for bleeding episodes, and, from the age of 8, suffered repeated hemorrhages into knees, elbows, wrists, ankles and fingers until a year prior to his first orthopaedic examination on November 11, 1951 At that time, Mr P was 34 years old, and although he had been afflicted with hemophilic arthropathies of the knees since childhood, he had never received any orthopaedic treatment aside from a single foot plate to unweight a painful callosity on the sole of his right foot

Constant pain in the knee joints had reached the point where daily doses of narcotics and sedatives were necessary to enable him to continue working as a salesman and to drive a car Recently, he had been placed on oral Cortisone in an attempt to eliminate both Demerol® and Codeine as far as possible

Describing his day, the patient stated that he arose at 6 A M and had to walk about with crutches or a cane for an hour in order to be able to leave the house at 7.15 A M Towards the end of the morning, he required Cortisone which provided relief until mid-afternoon when he repeated the dosage

During the past 4 years, his knee joints had become more and more painful He also suffered from a huge callosity in the metatarsal region of the right foot which was almost more disabling than his hemophilic arthropathies No one, however, had proposed to remove the callosity because of the patient's disease

Initial examination of knee joints revealed no swelling or evidence of fluid but marked crepitation and sensitivity to palpation and pressure The range of motion at the right knee was from AGE 150 degrees to AGF 100 degrees and, at the left knee, from AGE 140 degrees to AGF 80 degrees Dorsiflexion at the right ankle was restricted to 90 degrees, and elbows, though asymptomatic, showed mild deformity with restriction of extension more marked on the left side

Roentgen examination of the knees showed the characteristic destruction caused by advanced hemophilic arthropathy with very little articular cartilage left at any of the articular surfaces Juxta-articular bone cysts and coarse trabeculation were visualized at both knees

X-ray films also showed some involvement of the right ankle joint with thinning of the space between the tibia and astragalus, more marked on the fibular side

During a 22-day hospital admission, the large callosity under the second and third metatarsal heads of the right foot was easily and bloodlessly removed and three plantar warts beneath the callosity were excised A Lang-type celluloid foot plate was then manufactured to unweight the site of the callosity.

As the patient was dependent upon driving a car to earn his living, he preferred that treatment be limited to the left leg as he believed that major orthopaedic appliances for both lower extremities would make driving even more difficult Accordingly, the left leg was placed in traction suspension until a plaster of Paris model could be made of the leg for a light-weight hemophilia Hessing brace with Lofstrand spring joints at the knee A wedge cast was then applied which corrected the contracture at the knee to AGE 170 degrees After the new appliance was delivered, the patient was discharged from the hospital ambulatory without crutches and with no further pain on weight-bearing

At the end of the year, the patient demonstrated excellent results from wearing the hemophilia Hessing brace He walked better than he had in years with no recurrence of pain or swelling at the knee He was, however, still taking Cortisone every day

When next examined on February 23, 1952, Mr. P. reported that Cortisone medication had been discontinued a month earlier because of side effects of the drug and that he had since developed a great deal of pain in the right knee necessitating Codeine. While satisfied with the result of orthopaedic treatment of the left knee, he was still reluctant to submit to treatment for the right knee He was given a new attachment for further extension of the left knee consisting of a metal bridge that could be snapped on the Lofstrand spring joints and two metal plates with studs to be attached to the proximal and distal ends of the brace for application of elastics to be used daily for one hour.

From October 1952 to September 1953 the patient suffered no articular hemorrhages although several hematuria episodes required hospital admissions

When next seen on September 21, 1953, he had worn the well-fitting hemophilia Hessing brace for the left leg continuously, with no recurrence of pain in the knee He had, however, developed pain at the left ankle due to incongruence of brace and anatomical ankle joints which had occurred because the original brace had been made with the ankle joint attached to the foot plate to save weight At the time, it had not been sufficiently understood that incongruence of anatomical and brace axes might cause considerable joint pain in hemophilia.

The right knee had remained so painful that the patient continued to take regular doses of Codeine and Aspirin as well as Cortisone at certain intervals for exacerbation of pain

Examination showed that the left knee was completely painless with a range of motion from AGE 160 degrees to AGF 95 degrees. The right knee was painful with marked crepitation and tenderness on palpation and pressure over the medial articular space. Motion was possible from AGE 140 degrees to AGF 110

degrees It appeared that the only way to relieve symptoms and discontinue the daily use of narcotics and drugs was to fit the right leg with a hemophilia Hessing brace The patient, however, continued to resist this course of action

When examined more than a year later on October 21, 1954, the left knee had remained asymptomatic and showed good stability and a greater range of motion than could have been anticipated from the x-ray picture of advanced hemophilic arthropathy of long standing. The musculature of the leg had increased in strength and volume to such an extent that the brace had to be fitted with new leather cuffs Roentgen examination showed definite improvement in appearance of osseous structures in comparison with films taken 3 years earlier Calcium density was increased, trabeculation was better defined, and there had been no further destruction of articular space These findings clearly demonstrated the beneficial effects of the orthopaedic appliance that had been worn for 3 years

The right knee, however, had become increasingly painful with further destruction of articular cartilage and some additional decalcification of osseous structures At this point, the patient finally agreed to wear a second orthopaedic appliance

Immediate treatment for the right leg was precipitated by an acute locking of the knee joint, and the patient was admitted to the hospital on October 30, 1954 Examination of the joint revealed an advanced degree of hemophilic arthropathy and the knee was locked in extension of 140 degrees with extreme tenderness over the site of the internal semilunar cartilage There was local heat and discoloration of the skin over the medial aspect of the joint

Inasmuch as the acute condition of the right knee was not due to an articular hemorrhage but to the mechanics of locking by remnants of the internal semilunar cartilage, an intra-articular injection of 2 cc Hydrocortone was tried using the anterolateral approach next to the patellar ligament under cover of 3 cc 2 per cent Novocaine without Adrenalin Injection with a 24 gauge needle was painless and there was no bleeding. Pain disappeared and, after 24 hours, the right knee could be extended to AGE 165 degrees

By the end of a 28-day hospital admission, the patient had been fitted with his first hemophilia Hessing brace for the right leg with Lofstrand spring joints at the knee, as well as a reconstructed appliance for the left side to accommodate the increased musculature of the leg. A separate model of the right foot had also been made for a molded section to avoid pressure over painful callosities under the first and fifth metatarsal heads The single Hydrocortone injection into the right knee had proved most successful in relieving pain and increasing motion to the extent that the knee could be moved between AGE 175 degrees and AGF 120 degrees When discharged on November 27, 1954, the patient was completely ambulatory with his appliances

When examined again on June 29, 1955, knee joints were asymptomatic but the left ankle showed a mild degree of hemophilic arthropathy with small cystic areas in the astragalus and the os calcis, and some destruction of the articular cartilage of the subastragaloid articulation A plaster of Paris model of the left

leg was accordingly made for a new lower-leg and foot section for the hemophilia Hessing brace

At the most recent examination on May 24, 1956, recurrent symptoms at the left ankle were attributed to the fact that the hemophilia Hessing brace permitted considerable dorsiflexion of the ankle beyond 90 degrees while motion at the anatomical joint was possible to only slightly more than 90 degrees An ankle stop was therefore applied to the brace to limit dorsiflexion to 100 degrees This procedure, together with a lift on the heel of the left shoe to compensate for the resultant mild equinus position, eliminated all difficulties at the joint The patient was completely ambulatory with his two hemophilia Hessing braces which were entirely successful in keeping both knee joints asymptomatic

Case #5736

The orthopaedic history of Steven P is significant because of the remarkable rehabilitation of stance and locomotion after a single hospital admission for orthopaedic treatment

This patient, with a negative hemophilic ancestry, hemorrhaged profusely at circumcision, and experienced his first prolonged bleeding episode after a fence spike wound in the thigh at 5 years of age When he was 13, he was treated at a hospital in England for neurological symptoms with motor and sensory impairment in the left lower extremity, attributed to a possible hemorrhage into the spinal cord Motor symptoms disappeared, but sensory disturbances on the dorsum of the left leg, particularly around the knee, remained He also suffered a burn in this area from a hot water bottle. Gastro-intestinal and urologic bleeding during the next few years required hospital admissions and transfusions for each episode

After he arrived in the United States 3 years prior to his first orthopaedic examination, he was treated for a hemorrhage into the stomach Two years later, he was hospitalized for a fracture of the left tibia and treated with plaster of Paris and traction for eight weeks Although the fracture healed in the cast, the patient developed a permanent flexion contracture of the left knee Eight months before his first orthopaedic consultation, he was again hospitalized for cerebral hemorrhages Four months later, he was finally discharged from the hospital after the symptoms (unconsciousness, headaches) had subsided More recently, he suffered hemorrhages into left elbow and right ankle Frequently bedridden, he was bound to his cold-water walk-up apartment as he could rarely manipulate two flights of stairs At best, his crutch ambulation was very labored as he would not bear weight on the left leg

At the time of his first examination on November 8, 1951, the patient was 19 years of age. The right ankle exhibited symptoms of a recent hemorrhage, including painful motion, swelling, discoloration of the skin and local heat There was a flexion deformity at the left knee joint with very little active or passive motion The patellar reflex was not obtainable but this was not necessarily due to an absent reflex as the patella seemed more or less ankylosed to the femoral

condyles. The left arm, which had only recently required a transfusion, showed some enlargement and restriction of extension at the elbow

Roentgen examination revealed fairly extensive decalcification of osseous structures of the left knee, irregularity of femoral surfaces and almost complete obliteration of the postpatellar joint space X-rays of the left ankle showed moderate decalcification of all osseous structures.

After a single transfusion for rectal hemorrhages, the patient was hospitalized on November 14, 1951 for 33 days for a flexion contracture of 135 degrees at the left knee During this admission, he was fitted for a hemophilia Hessing brace with Lofstrand spring joints at the knee and a sandal for the foot. At the time of discharge, the knee joint showed a range of motion from AGE 150 degrees to AGF 110 degrees, the patient walked comfortably and was able to manipulate stairs This hospital admission was complicated by the emergency extraction of an abscessed lower left first molar, followed by bleeding from the tooth bed for several days and necessitating transfusions of whole blood and plasma

By February 1952, Steven had made rapid progress without further treatment and there had been no recurrence of hemorrhages. Providing comfortable ambulation without a cane, and allowing the patient to walk up and down the two flights of stairs to his flat, the orthopaedic appliance was well tolerated and the Lofstrand spring joints at the knee had successfully increased extension to 175 degrees

Two months later, he was admitted to the hospital for extraction of a right upper molar and remained there for 62 days Orthopaedic examination during this period showed that 10 degrees of extension at the left knee joint had been lost Although the patient made a complete recovery from dental surgery, considerable extension at the left knee was forfeited as he had not worn the hemophilia Hessing brace while bedridden After the appliance was reapplied, elastics were added to obtain complete extension

A routine examination in March 1953, showed that extension at the left knee had again reached 165 degrees A year later, the patient walked so well that he had lost weight and the condition of the left knee was as good as could have been expected The joint was held rigid in extension of 170 degrees with some posterior subluxation of the tibia in relation to the long axis of the femur Roentgen examination showed slightly better calcification of the skeleton of the knee, absence of soft tissue swelling, and no evidence of further deterioration of articular space

When last seen in July 1954, the patient was fitted with a new light-weight hemophilia Hessing brace without knee joint, and was considered completely rehabilitated from the orthopaedic viewpoint

Case #8058

Martin J, age 26, serves as one of the more graphic examples of complete rehabilitation of an adult patient with a severe hemophilic arthropathy This patient, whose uncle was the only other member of the family or ancestry known

to be so afflicted, was born with a large hematoma on his head, and classified as a hemophiliac at that time

Aside from joint bleeding, he suffered only one serious hemorrhage during his early years when the upper lip was cut in a fall. After the child had bled for a month, hemorrhages ended *spontaneously. At 5 years of age, when two* baby teeth were removed, and again at age 11, when the left thigh bone was broken, there were no complications

At age 24, the left knee joint became involved following a simple contusion, thereafter elbows and the right knee were affected The only dangerous episode other than kidney hemorrhages during early adulthood occurred when the patient was 25 years old and suffered a hemorrhage under the tongue. As swelling threatened the air supply, the patient required immediate hospitalization for transfusions.

For the 2 years preceding our first orthopaedic examination, the disability at the left knee had become increasingly acute Despite 6 weeks of hospital treatment in 1955, the patient remained crutch ambulatory and found himself unable to continue at his clerical job.

In this condition, the patient was finally hospitalized on February 2, 1956 for orthopaedic treatment of the painful flexion deformity at the left knee Although elbows and the right knee were also involved, the patient requested that treatment be limited to the left knee

First orthopaedic examination on February 3, 1956 revealed considerable enlargement of the left knee without ballottement of the patella. Extension was restricted to 140 degrees and flexion to 75 degrees, with a valgus deformity of 10 degrees. The musculature of the entire leg was atrophic The right knee moved from 180 degrees to 95 degrees and showed less valgus deformity than on the left side.

Roentgen examination of the left knee presented a complete picture of severe hemophilic arthropathy although generalized decalcification was less extensive than anticipated on the basis of clinical findings Femoral and tibial condyles were blown up All articular surfaces were destroyed with complete breakdown into juxta-articular cysts and cavities. Because of the recent origin of the hemophilic arthropathy, the diaphysis of femur and tibia and the fibula proper were better developed than is usual for this degree of articular destruction and deformity of condyles The patella, however, showed squaring off of its distal pole demonstrating involvement with hemophilic arthropathy prior to skeletal maturity. The tunnel view showed almost complete absence of articular space The lateral exposure indicated considerable thickening of soft tissue structures about the knee compatible with a more acute phase of joint disease.

Films of the right knee exhibited better calcium density of all osseous structures and clear trabeculation of metaphyses and epiphyses Nevertheless, there were many juxta-articular cysts, particularly in the medial condyle of femur and tibia Some of these, indicating repair, may have been filled in with bone. The marked deformity of the patella *indicated early involvement.* There was no evidence of soft tissue thickening around the joint.

The right elbow showed a mild degree of pathology with enlargement of the head of the radius and some deformity of articular surfaces Calcium density was almost normal and the articular space was fairly well preserved The left elbow revealed a more marked degree of involvement, with some valgus deformity, enlargement and deformity of both the radial head and the coronoid process of the ulna. Articular space was markedly diminished and there were small juxta-articular cysts

Immediately after admission to the hospital, plaster of Paris models were made of both legs from toes to below the groin to be used in the manufacture of a hemophilia Hessing brace for the left leg with Lofstrand spring joints at the knee, simple ankle joint and sandal, and for a smaller protective appliance for the right knee The latter was indicated, despite the fact that the right knee was clinically asymptomatic, because of the considerable destruction of articular surfaces visualized in x-ray films

As successful correction of the deformity at the left knee could readily be anticipated, this was one of the rare cases in which a plaster model for a major orthopaedic appliance could be taken prior to application of a corrective cast Directly thereafter, a Quengel cast hip spica was applied for correction of the flexion deformity of the left knee and well tolerated When the cast was removed, there was no pain, swelling, or local heat at the knee joint and extension had been increased from 140 degrees to 160 degrees

After an initial fitting of the new hemophilia Hessing brace, a wedge cast was applied from toes to below the groin to obtain extension beyond 160 degrees No attempt was made to correct the 10 degree valgus deformity resulting from overgrowth of the medial femoral and tibial condyles earlier in life The patient was discharged at the end of a 29-day hospital admission wearing a hemophilia Hessing brace on the left leg and a Hewitt elastic knee brace on the right The flexion contracture of the left knee had been corrected and the joint was asymptomatic. For the first time in almost 2 years, Mr J was without pain on weight-bearing and completely ambulatory without crutches

At a follow-up examination on May 19, 1956 the patient appeared in fine spirits. Free of all disabling symptoms, he had become ambulatory without canes and able to lead a productive life

Case #6975 (See Figures 15, 20 and 22)

Alan W was 45 years of age at the time of our first orthopaedic examination, having suffered from hemophilia since infancy His family background included a *nephew who was also a known hemophiliac Recurrent attacks of pain and* bleeding for a period of 8 years had seriously interfered with his occupation as a taxicab driver and had reached the point where the patient was able to work for only 4 months each year Pain was most marked when the patient arose from a sitting position

At the first orthopaedic examination on April 5, 1954, the patient appeared in moderately good general health He walked with a marked right-sided limp, holding the leg in external rotation The right ankle was swollen and quite painful

with marked restriction of motion in all directions and loud crepitation on passive movement There was only *mild involvement of the left foot and ankle* Both knee joints exhibited conspicuous hemophilic arthropathies with flexion contractures There was some atrophy of the musculature of the right lower-leg in comparison with the left side. Elbow joints had also been affected by hemorrhages.

Roentgen examination revealed severe bilateral knee involvement far exceeding clinical findings Destruction of all articular cartilage had reduced articular space to a minimum with considerable deformity of articular surfaces. The picture was dominated by a large number of *juxta-articular cysts, particularly* in the medial femoral and tibial condyles where they were of considerable size. There was also sclerosis at articular margins Productive changes with osteophyte *formation, rarely seen in hemophilic arthropathy,* were noted at the posterior aspect of the femoral condyles. The osteophyte on the left femur had the configuration of an osteochondroma The left patella showed little deformity but marginal lipping and small cysts. The right patella showed considerable enlargement of its anteroposterior diameter, with one large and several small cysts

At the right ankle, cartilage was almost completely destroyed leaving very little articular space. The juxta-articular portions, particularly at the astragalus and the external malleolus, were full of small cysts with some productive changes and *lipping anteriorly.*

The structure of the left ankle was better preserved than that of the right, but also exhibited diminished articular space and deformity with several juxta-*articular cysts*

There was some generalized decalcification of the osseous structures of both feet and thickening of the midshaft of the third metatarsal but no signs of hemophilic arthropathy

In contrast to x-ray evidence of advanced joint destruction, both knee joints showed surprisingly little functional disability. The hemophilic arthropathy of the right ankle presented the main problem for rehabilitation This called for an orthopaedic appliance that would permit use of the right foot on the brake of the taxi without trauma to the severely diseased ankle joint. On April 12, 1954 a plaster of Paris model was made of the right leg for a lower-leg Hessing brace with milled ankle joint incorporating a stop for dorsiflexion to eliminate impact of the diseased articular surfaces of ankle mortise and astragalus when the foot was applied to the brake. After delivery on May 11, 1954, beneficial effects of the Hessing brace were immediately apparent and the patient was able to continue at his job without significant disability through December 1954.

For the next 7 months, the patient's condition was aggravated by pain in the left hip, frequent small hemorrhages into wrists caused by weight-bearing on the arms when getting in and out of his cab, and considerable pain and swelling at the right lower-leg due to a brief period of ambulation without his orthopaedic appliance At that time, he was instructed to wear his brace continually. Mainly, however, recurrent pain, swelling and stiffness at the elbows prevented the pa-

tient from working in his cab. He stated that as soon as he attempted to drive, his elbows became painful and he was forced to rest at home.

Examination on July 12, 1955 showed deformities of the elbows, with marked restriction of motion in both directions X-rays revealed advanced hemophilic arthropathy bilaterally, with enlargement of the radial head and a very large number of juxta-articular cysts This condition did not appear to be amenable to active treatment.

The patient had not worn his right lower-leg Hessing brace, with which the ankle joint had remained asymptomatic, because of a fracture of the second metatarsal 2 weeks earlier. X-ray films showed the fracture healing in good

already visualized

August 8, 1956 after a serious fall which
mination revealed massive discoloration
x from the recent hematoma with swell-
tissue most marked over the sternum
hages into the joints with restriction of
recovering from these injuries, the pa-
edic treatment at the time

the patient reported his narcotic addic-
rge doses of morphine for many years,
e withdrawal of the drug and was once
he believed, did not know of his addic-
at complete withdrawal, he had placed
o had reduced his injections with great
h agent of the narcotic squad, the drug
ent could not tolerate the rate of with-
usea and vomiting It was in this con-
nd elbows when he slipped and fell in

brace was outstandingly successful in
a recurrence of hemorrhages into the
ed him from continuing at his occupa-

rald E was 54 at the time of his first
patient revealed a hemophilic family
d of the disease in middle age
ed at circumcision and lasted 4 weeks
ng in infancy, the patient's condition
0 years of age. In addition to kidney
bleeding in all parts of the body, with
f the tongue that tube feeding through
the nose became necessary. While internal hemorrhages were so severe that the

patient was bedridden for months at a time, articular hemorrhages during that period caused no great damage.

This patient is one of the few recorded cases to survive an appendectomy or any other type of extensive surgery prior to the use of antihemophilic plasma. Sustained at age 26, the operation was followed by severe hemorrhages requiring a 3 months' hospital stay Complete recovery was possible only after a full year

At age 30, the patient suffered his first serious articular hemorrhage following an injury to the left knee After aspiration of blood and application of a cast to the leg, the patient was on bed rest for 6 months before ambulation could be resumed

A year later, severe hemorrhages into the right elbow caused a flexion contracture, permitting only restricted use of the arm and hand.

During the years from 40 to 52, Mr E was bedridden 50 per cent of the time with recurrent hemorrhages, swelling and great pain at the right knee With aspiration of blood on several occasions and treatment of the joint by a succession of casts, the hemorrhages persisted and the patient was barely able to walk with crutches. In addition to hemophilic involvement of knees, ankles and elbows, the patient's general condition was complicated in November 1953, by a cerebral vascular accident resulting in a right hemiplegia At the time of his first orthopaedic consultation on May 6, 1954, he had been hospitalized for medical treatment of his critical cardiovascular status, and was totally disabled. Although the tendency toward hypertension and bleeding was in the foreground, most of the sequelae of the hemiplegia had subsided but Mr. E was unable to handle a crutch or cane with his right hand and had not walked in six months.

Examination revealed swelling and pain at the right knee with local heat and tenderness most marked over the medial aspect. A considerable deformity was exhibited with AGE 140 degrees and lateral and posterior subluxation of the tibia

Although the left knee joint was painless, it showed a marked deformity with a range of motion from AGE 165 degrees to AGF 110 degrees Due to longstanding postero-lateral subluxation of the tibia, the medial condyle of the femur was quite prominent.

The right elbow showed a considerable restriction of motion due to involvement with hemophilic arthropathy of long standing with a range of motion from AGE 140 degrees to AGF 100 degrees. This joint could not tolerate the strain of crutch or cane ambulation

Clinical examination of ankles revealed some tenderness with restriction of motion

Roentgen examination revealed advanced involvement with hemophilic arthropathy of recent origin at the right knee joint with increased soft tissue density and generalized decalcification Juxta-articular cysts were present throughout the medial femoral and tibial condyles and in the patella. Bone destruction was most noticeable at the medial margin of the femoral condyles, where severe pain was apparent on clinical observation.

The left knee joint showed quite marked deformity with posterior and lateral displacement of the tibia in relation to the femur. There was generalized decalcification of all osseous structures with large cystic areas close to the articular surface of the femoral condyles, more marked on the medial side The patella exhibited some deformity and large cysts were observed Articular space between the patella and femur was almost non-existent Productive changes were most obvious between the medial femoral and tibial condyles. With no suggestion of an active process, the marked deformity was due to long-standing hemophilic arthropathy

At the right elbow, there was a moderate degree of hemophilic arthropathy of long standing Calcium density of the osseous structures was almost as good as at the normal left side All articular surfaces showed destruction and deformity with juxta-articular cysts most marked at the ulnar aspect of the joint There were also cystic areas in the head of the radius

The ankles showed generalized decalcification of mild degree on the right side, attributable to the 1953 hemiplegia, and thinning of articular cartilage of the left ankle joint with slight defects in the articular surface of the astragalus and minimal juxta-articular cyst formation, particularly in the distal end of the tibia

It was recognized that any attempt at orthopaedic rehabilitation entailed considerable risk for a person of Mr E's age and cardiovascular condition Nevertheless, after he was medically stabilized, he was transferred by ambulance from another hospital on May 8, 1956, for orthopaedic treatment of the acute and very painful hemophilic arthropathy of the right knee and for lesser involvement of left knee, right elbow and right ankle.

After complete immobilization of the right leg, pain and local heat subsided As soon as extension of the right knee had reached 165 degrees, plaster of Paris models were made of both legs to be used in the manufacture of hemophilia Hessing braces with Lofstrand spring joints at the knee and milled ankle joints The right knee was then immobilized in a maintenance cast In addition, a plaster of Paris model was made of the right arm from the deltoid muscle to above the wrist for a fiberglass brace with lock joint at the elbow to prevent hemorrhages from the use of crutches or canes

After all appliances were fitted and delivered, walking exercises between parallel bars were started, and the patient was soon able to stand unsupported and walk quite well pushing the wheelchair ahead of him Mild symptoms of angina were the only complications during this hospital admission which lasted 32 days At the time of his discharge on June 9, 1954, the patient's rehabilitation far surpassed expectations *He had become ambulatory with two canes and* was capable of manipulating stairs Flexion contractures at the knees were completely corrected Blood pressure was lower than upon admission to the hospital and his general condition had improved

Letters received thereafter testified to the renewed vigor and confidence of the patient Walking without crutches or canes, he was able to return to work and travel, restricted only by his heart condition He continued to enjoy complete

orthopaedic rehabilitation until he suffered a final cerebral hemorrhage from which he died in January 1956

Case #5747

Carlos M was 13 years old when his family moved from their home in Venezuela to New York City in November 1951 for the sole purpose of obtaining treatment for their son's orthopaedic disability. Diagnosis of hemophilia in Carlos' case was made at 10 months of age and the family history included two uncles who had died of the disease.

Joint involvement for the patient began at age 6 with the left knee, followed by hemophilic arthropathy of the right knee at age 10, and subsequent involvement of elbows, ankles, left hip and right shoulder. As the patient had also sufferred hemorrhages from the throat and kidneys, he had come to New York City once before for hematological examination and treatment but did not require transfusions at the time. On the orthopaedic level, correction of a flexion deformity of the left knee was unsuccessfully attempted first by traction and then by means of a night splint

Because of severe flexion deformities at both knee joints, the patient had been wheelchair ambulatory and totally unable to stand or walk for the 3 years prior to our first examination. Sporadic formal education had, of necessity, been carried on at home.

The first orthopaedic examination on November 16, 1951 revealed that despite flexion contractures at both knee joints, the patient's musculature was surprisingly well-preserved due to adept handling of his wheelchair. The left knee joint appeared in worse condition than the right, both clinically and roentgenologically Range of motion was limited from AGE 125 degrees to AGF 95 degrees and the patella was so deep seated between the enlarged femoral condyles that it could not be palpated The right knee joint moved with greater ease between AGE 120 degrees and AGF 40 degrees and the musculature of the right leg was considerably stronger than that of the left.

X-ray films taken on November 20, 1951 showed quite marked decalcification at the knees with some flattening of femoral condyles and slight irregularity of the posterior border of the patella All changes were more extensive on the left side At the ankles, there was rather marked decalcification with the talus in valgus position. There was considerable decalcification of osseous structures of the feet with bilateral pes cavus and metatarsus adductus of moderate degree. Films of the elbows revealed mild destructive changes at the articular surfaces of the humerus

Despite bilateral knee involvement, it appeared psychologically judicious from the patient's standpoint to treat first that joint which would yield more rapidly to corrective forces. For this reason, the right knee was selected

Accordingly, the patient was hospitalized on November 28, 1951 for application of a Quengel cast hip spica to the right leg. After 6 days, he was discharged wheelchair ambulatory with the Quengel cast All necessary hospital equipment was set up in the family's apartment in New York and the patient's private nurse retained

During ambulatory treatment handled either at home or by means of several ambulance visits to the hospital from December 4, 1951 to February 28, 1952, correction by the Quengel method obtained extension to 155 degrees at the right knee After a plaster of Paris model was made of the right leg to be used for a hemophilia Hessing brace with Lofstrand spring joints at the knee, a second Quengel cast was applied until the brace was delivered and later fitted with elastics to be used periodically for additional extension at the knee

As treatment had proceeded smoothly to this point, it became advisable to initiate correction of the left and more severely damaged knee joint by means of a Quengel cast, followed by application of a wedge cast with the knee held in extension of 150 degrees A series of wedgings obtained extension to 160 degrees After the cast was removed, a plaster of Paris model was made of the left leg for a hemophilia Hessing brace with Lofstrand spring joints, milled ankle joint and sandal A maintenance cast was then applied holding the left knee at AGE 160 degrees until the brace could be fitted and delivered Responding to treatment much more readily than anticipated, the left knee rapidly reached and maintained extension to 165 degrees and remained free of pain throughout the entire corrective procedure

The right knee, however, underwent a number of setbacks and by February 1952 had suffered several episodes of pain and swelling A considerable degree of extension was lost and the knee required temporary immobilization in the brace, with Lofstrand spring joints locked by set screws at 140 degrees At one point, an effort was made to improve the painful condition of the joint by a single injection of 1 cc (25 mg) Hydrocortone Although this produced no untoward effects, it did not help the knee

Further examination on May 1, 1952 revealed excellent progress at the left knee in spite of x-ray evidence of much greater joint destruction than on the right side The joint was completely free of pain and swelling, maintaining extension to 170 degrees in the hemophilia Hessing brace

The right knee joint remained painful with extension limited to 145 degrees Pain originated from the medial aspect where there was some swelling and well localized tenderness on pressure over the femoral condyle Any attempt to further extend the knee caused additional pain and muscle spasm X-rays taken at this time showed an area of irregular destruction of one section of the articular surface of the medial condyle of the femur, probably the site of an active lesion The patient was advised to wear his hemophilia Hessing brace continuously and to make no effort to flex or extend the knee, as the joint required rest until inflammatory changes had subsided By May 17, 1952 acute inflammatory changes in the joint had completely subsided but extension was limited to 130 degrees A wedge cast was applied to the right leg, obtaining AGE 155 degrees at the knee Elastics were then added to the hemophilia Hessing brace for further extension

Corrective treatment and rehabilitation during these first nine months were impeded not only by the inflammatory reaction of the right knee joint but also by general factors such as fast growth during adolescence necessitating frequent adjustment and repair of appliances Progress was further retarded by bleeding

into both elbows and the left shoulder as well as by kidney and abdominal hemorrhages. Sinusitis was an additional irritant. Recurrent headaches and twitches in the left arm, suggestive of a mild form of epilepsy, also served to complicate the boy's condition

Nevertheless, by September 1952, the right knee was sufficiently improved, with extension to 155 degrees, to allow the patient to stand and walk, aided by two canes or a walker, for the first time in 4 years. Attempting to control the continuing pain in the right knee joint, small doses of oral Cortone were administered. This medication, however, did not agree with the patient and was discontinued

By January 1953, the right knee appeared considerably better as a result of the protection offered by the well-fitting hemophilia Hessing brace. The joint was no longer swollen and there was no pain over the medial articular space.

At about this time, the painful condition of the elbow joints had become acute as a result of crutch ambulation Examination of both arms on January 4, 1953 revealed that restriction of pronation to less than 50 per cent of the normal range of motion on both sides was probably the main cause of pain. The arms showed residual swelling and slight discoloration of skin throughout the volar and medial aspect. Extension was possible to 140 degrees at the left elbow and to 150 degrees at the right, with no restriction of flexion. The musculature was in good condition though slightly weaker on the left side

X-ray films of elbow joints, dated January 8, 1953, showed cartilage and juxta-articular bone destruction at the joint between the right humerus and ulna A slight deformity of the radial head and decalcification of all osseous structures were also noted. The left elbow showed slight decalcification with a minimal deformity of the radial head and a small amount of cartilage destruction along the coronoid process of the ulna.

Plaster of Paris models were made of both arms to be used in the manufacture of fiberglass appliances for support and protection of the elbows. The brace for the left elbow consisted of a jointless protective splint while that for the right elbow had double bars and a joint at the elbow that could be locked at 130 degrees of extension and at 90 degrees of flexion. The patient also received Sterling Bunnell's "active splints" for increasing pronation of the forearms, and a pair of forearm crutches

By March 1953, the boy was able to stand and walk with very little support, preferring regular crutches to cane-gliders or forearm crutches By May 1953, he was in excellent general condition, completely crutch ambulatory and able to stand unsupported

The left knee was fully extended and free of swelling and pain, exhibiting, however, a pronounced valgus tendency The right knee was painless and extended to 165 degrees, having been at that angle for several months. Both elbows had become asymptomatic

During these 17 months of treatment from November 1951 to May 1953, which coincided with a period of rapid growth for the patient, avoidance of fatigue, so hazardous in treatment of a hemophiliac, was essential Although it

was difficult to keep the patient from attending too many movies or watching too much television, his behavior was generally cooperative and helpful

After returning to Venezuela for 17 months, Carlos came back to New York City for examination and for new orthopaedic appliances as his braces had become too small Demonstrating an unusual degree of rehabilitation upon examination on October 26, 1954, he was able to stand and walk quite well without appliances and crutches Although he was without pain, he tired quite easily and preferred ambulation with crutches Examination revealed that the left knee was completely straight and the valgus position less marked than previously noted, while the right knee still showed moderate enlargement and restriction of extension to 165 degrees There was some limitation of motion at both elbows but no pain The musculature of both arms was excellent and hands had developed strength

A series of x-rays taken at this time showed a considerable recalcification, much improved trabeculation and no progression of hemophilic arthropathies in comparison with previous films

Rehabilitation had progressed to the point where the patient required lighter and smaller orthopaedic appliances Plaster of Paris models were made of the right leg for a lighter hemophilia Hessing brace with Lofstrand spring joints at the knee, and of the left leg for a fiberglass knee splint with simple joint at the knee and a Whitman plate for the left foot As the left leg was ½ inch shorter than the right, a compensatory ½ inch lift was ordered for the heel of the left shoe to correct the pelvic obliquity and postural scoliosis

All appliances were fitted and delivered and the patient left for Venezuela in December 1954 with instructions to return in 2 years for a check on orthopaedic appliances A letter, received in March 1956, reported that the patient was continuing at the same high level of rehabilitation

Case #6287

For 15 years, Peter M had been denied all opportunity for the kind of treatment that might have obviated the development of extensive articular damage This had resulted from his mother's apprehensiveness following a transfusion that allegedly had caused a paralysis during her son's infancy Even after many years, it took a great deal of persuasion to obtain permission for a second transfusion in November 1952, which was extremely necessary and highly successful During a preliminary interview with the boy's parents, it became apparent that they were still doubtful about the advisability of orthopaedic treatment

At the time of his first orthopaedic consultation on December 2, 1952, Peter was 15 years of age, wheelchair ambulatory and able to move about by pushing himself with his feet on the ground Although he was a good swimmer, he had never been able to stand or walk With an ancestry that was negative for hemophilia, he had suffered from the disease since infancy and by early adolescence was afflicted with hemophilic involvement of knees, elbows and the right ankle

A complete examination showed markedly atrophic musculature of the legs, considerable deformity of knee joints with flexion contractures, valgus position

and external rotation of the lower-legs. Posterior subluxation of the tibia in relation to the long axis of the femur was more marked on the right side. The right knee showed a range of motion from AGE 100 degrees to AGF 40 degrees while the left knee moved from AGE 110 degrees to AGF 65 degrees Dorsiflexion of both ankles was limited to 110 degrees Motion at the elbow joints ranged from AGE 150 degrees to AGF 55 degrees on the right side and from AGE 135 degrees to AGF 55 degrees on the left.

Roentgen examination revealed advanced hemophilic arthropathy of knees, right ankle and left elbow, moderate involvement of the right elbow, and, at the left ankle fairly good preservation of the joint but considerable decalcification of all osseous structures Films of knee joints showed the usual characteristics of advanced hemophilic involvement with blown up femoral and tibial condyles in contrast to atrophic femoral shafts and with deformity and cyst formation of the patella Articular space was better preserved on the left side and the flexion contracture was more marked on the right.

As these deformities were too marked and too rigid to permit more than partial correction at any one hospitalization without the risk of fatigue-induced hemorrhages and possible damage to blood vessels and nerves, orthopaedic treatment had to be planned over a 3-year period. Furthermore, as the patient was only 15 years old, it was felt that mild corrective forces introduced by orthopaedic appliances could influence the development of osseous structures during the subsequent 3 or 4 years and thereby achieve the optimum weight-bearing alignment.

The original plan called for one month of hospitalization each year The first admission, however, lasted 85 days from February 2, 1953 to April 28, 1953 because of the large number of Quengel and wedge casts required to obtain even minimal correction of long-standing hemophilic deformities By the time a pair of hemophilia Hessing braces with Lofstrand spring joints at the knee was finally delivered, the left knee was held in extension of 165 degrees and the equinus tendency of the left foot had been gradually corrected by elastics The right knee had offered much greater resistance and it was not until nearly the end of the 3-month hospital admission that the joint could be extended to 145 degrees and the appliance for the right leg fitted Thereupon, the patient received instructions in ambulation between parallel bars, and, as maintenance of the upright position was extremely difficult, the ankle joint of the right hemophilia Hessing brace was locked at 100 degrees, and the Lofstrand knee joints set at 145 degrees to provide greater stability

As the patient had never stood or walked before, orthopaedic correction could not achieve even the beginnings of crutch ambulation at this time. Nevertheless, maintenance of the upright position and ambulation between parallel bars were measures of marked progress from the patient's original condition

When the boy was next examined on April 21, 1954, he appeared more mature and was able to stand and walk to some degree with Lofstrand crutches. His posture was characterized by a considerable lack of extension at the right knee and to some extent at the left, and hindered by a deficiency of dorsiflexion at

the ankles These articular restrictions made it impossible for him to stand with hips extended. Roentgen examination showed AGE 150 degrees at the left knee and AGE 140 degrees at the right knee with better calcification and some clearing of structural details

The second hospital admission for further correction of flexion and valgus deformities of the knee joints was begun on April 24, 1954. As deformities had remained marked, another large Quengel cast double hip spica had to be used Quengel correction of the left knee proceeded quickly and was started at the right knee after the range of subluxation hinges for correction of posterior subluxation of the tibia had been nearly exhausted After construction of new hemophilia Hessing braces, the patient started to walk once again between parallel bars A pair of fiberglass knee braces, to be worn for swimming, was also made to plaster of Paris models and the patient was discharged crutch ambulatory after a 31-day admission Plaster of Paris treatment during this period had increased extension at both knees by 20 degrees, and part of the valgus deformity of the right leg had been corrected A single episode of hematuria, requiring two transfusions with antihemophilic plasma, did not retard corrective procedures during this hospitalization

The third hospital admission from April 11, 1955 to May 6, 1955 achieved a higher level of rehabilitation In the interim, the patient's condition had been complicated by an acute hematuria episode requiring transfusions with a combination of whole blood and plasma during the summer of 1954, and multiple hemorrhages into the right knee in September 1954 Since then, there had been no further hemophilic symptoms The musculature of both legs had gained in strength and volume Knee joints had remained asymptomatic Motion at the right knee ranged from AGE 150 degrees to AGF 90 degrees with 15 degrees of valgus deformity due to overgrowth of the medial femoral condyle, and at the left knee from AGE 155 degrees to AGF 115 degrees with a valgus deformity of 12 degrees Dorsiflexion of feet had improved to 105 degrees on the right side and to 80 degrees on the left Hip joints were freely movable with no trace of flexion. While the boy spent a good deal of time in his wheelchair, locomotion had been greatly improved He was able to stand and walk without crutches, and hardly used a cane

Roentgen examination of the knee joints revealed a remarkable amount of recalcification of all osseous structures and no further bone destruction There was also much improvement of calcium density and trabeculation at feet and ankles, and all osseous structures were better developed

An acute hemorrhage into the volar aspect of the right forearm, extending to the dorsum of the hand with residual flexion contracture of the right fourth finger, did not interfere with the course of rehabilitation of the lower extremities Treatment of the arm, however, was urgently needed and immediately upon admission to the hospital, the right hand and the forearm were splinted and treated with ice Residual infiltration and induration of soft tissues on the volar aspect of the wrist, and restriction of complete extension of the fourth finger were treated with a dorsal aluminum splint applied to the forearm, with an aluminum

post for attachment of elastic traction to correct the flexion contracture of the finger After this was accomplished, the splint was discarded

Furthering rehabilitation of the lower extremities, wedge casts were applied to both legs holding the left knee in extension of 150 degrees and the right at 145 degrees. After a series of wedgings on both sides, extension at the knees had reached 170 degrees, but no diminution of the bilateral valgus deformity had been obtained. It appeared that orthopaedic correction had proceeded as far as possible and that residual deformities of the lower extremities would be permanent. Knees were rigidly splinted until a new pair of hemophilia Hessing braces could be delivered With these well-fitting appliances, the patient's posture was greatly improved Standing erect with very little residual flexion at the knees, he began to walk without crutches.

When seen again on December 27, 1955, the boy had grown considerably and at the same time had lost weight. As a result, his legs moved up and down in his braces and the anatomical and brace joint axes were no longer congruent This had led to a series of hemorrhages into the right knee and one into the left knee, between May and November 1955. Also during this period, he had been hospitalized for several days with an intestinal hemorrhage, and had suffered a hemarthros at the left wrist Examination further revealed that the patient's posture, though tending slightly toward flexion at the hips, was satisfactory The right knee joint had remained extended to 170 degrees with a valgus deformity of 10 degrees and the left knee was extended to 165 degrees with a 10 degree valgus deformity. External rotation of the right lower-leg was more marked

Inasmuch as no further correction of deformities was considered feasible, plaster of Paris models were taken for new appliances designed to eliminate hemorrhages into knee joints and irritation of ankles by establishing correct axes alignment It was planned that the patient should be briefly hospitalized on January 2, 1956 for fitting and delivery of his new hemophilia Hessing braces This admission, however, lasted 21 days due to an acute hemorrhage into the left groin and hip region which had started 48 hours prior to hospitalization. After the bleeding had been controlled by rest and transfusions, development of a flexion contracture was avoided by proper positioning in a fracture bed. As a result of this extensive hemorrhage, which was followed by marked secondary anemia, the patient lost so much weight that fitting of the new appliances presented a problem As an alternative to making new plaster of Paris models for alteration of the braces to conform to the legs, the braces were fitted with a filler of foam rubber to be used until the boy regained his normal weight. He was discharged from the hospital on January 23, 1956, walking very well, both with and without crutches, and holding himself more erect than ever before. The most recent report from the patient, dated June 3, 1956, indicated that he was extremely comfortable with his new orthopaedic appliances.

Case #7513 (See Figure 30)

A malunited fracture of the left femur had reduced John B., age 9, to a state of total disability at the time of his first orthopaedic examination on March 14,

1955 In addition, the child urgently required psychiatric treatment for behavior problems stemming from a difficult family situation

With a background that was negative for hemophilia, the child exhibited abnormal bleeding tendencies directly after circumcision Multiple subcutaneous hemorrhages starting at 4 months of age were followed by frequent hemorrhages into the left knee causing joint deformity with moderate restriction of extension and flexion. The hemophilic joint had not been braced and the child had attended school whenever he was not confined to bed

In September 1953, the patient was hospitalized for 5 weeks with a severe hemorrhage into the volar aspect of the left forearm that led to the development of a claw hand. A banjo splint, applied for correction of the flexion contractures of the long fingers, was unsuccessful and the patient finally refused to wear it

In December 1954, the patient suffered a severe fall resulting in a fractured left femur Plaster of Paris treatment during a 15-week hospital admission was so inadequate that when the last cast was removed early in March 1955, the knee was stiff, the fracture improperly healed, and the patient unable to walk Furthermore, a hemorrhage had occurred into the extremity during the time the leg was supposedly immobilized in a cast

Clinical examination of the left leg showed enlargement of the thigh due partly to callus formation at the fracture site and partly to an organized or calcified hematoma in the soft tissues The knee joint was rigid in extension of 150 degrees but otherwise normal

The undersized left forearm and hand were considerably atrophied with the forearm almost rigid in pronation, the wrist free and the proximal interphalangeal articulations of the four long fingers held in extreme flexion With the wrist held in maximum volar flexion, there was considerable extension of the fingers

Roentgen examination of the left leg revealed an oblique fracture throughout the mid-portion of the shaft of the femur united with bony callus The fragments were in marked angulation with a moderate degree of overriding A tremendous amount of soft tissue swelling was visualized involving the entire left thigh Much of the soft tissue between the angulated fragments showed increased density and areas of calcification The knee showed a rather advanced state of hemophilic arthropathy with decalcification, destruction of cartilage and juxta-articular cysts with enlarged epiphyses There were also signs of hemophilic arthropathy at the ankle

X-ray films of the left elbow showed a moderate hemophilic arthropathy, with the epiphyses appearing much larger than those of the normal right elbow

Although prognosis for orthopaedic rehabilitation was uncertain because of the severity of the original involvement of the left forearm, left knee and ankle, and the malunited fracture of the left femur, the patient was hospitalized for 13 days from March 21, 1955 to April 3, 1955, and treatment for flexion contractures of the left hand and fingers was begun with a Quengel cast This was followed by application of a second cast with elastic traction for the second, third and fourth fingers

While surgical correction of the marked deformity of the left leg was im-

possible because of massive callus formation and hemophilia, some rehabilitation of locomotion was feasible. As a major orthopaedic appliance was required for protected weight-bearing, a plaster of Paris model was made of the extremity for a hemophilia Hessing brace with Loftstrand spring joints at the knee. Immediately upon delivery of this appliance, the patient became cane ambulatory and able to bear weight upon the left leg without pain.

This single hospital admission was followed by 3 months of concentrated treatment for the left hand. Extension of the fingers began to improve steadily and the patient was able to start motion at some of the terminal articulations The Quengel cast was replaced by a maintenance cast holding the wrist in dorsiflexion of 25 degrees and the fingers in maximum extension of proximal interphalangeal articulations. After additional Quengel treatment, the patient received a fiberglass appliance designed with an aluminum plate to which the four fingers were attached for elastic traction on the middle and terminal phalanges.

When last examined on August 8, 1955, the patient continued to be cane ambulatory without pain, the deformity from the malunited femoral shaft structure was less marked and function of the left leg was sufficiently restored to permit the patient to swim.

This degree of rehabilitation, together with the scheduling of psychiatric treatment, offered a more optimistic future for the boy than could have been anticipated.

Case #3587 (See Figures 9 and 36)

Christopher L., the first patient treated in this series, came for his initial orthopaedic consultation in 1946, at the age of 4 This was also the first case in which the Quengel method of orthopaedic rehabilitation was used for contractures resulting from hemophilic arthropathies In a sense, the recounting of this history details the progress made in the orthopaedic treatment of hemophilic sufferers in the past 10 years.

With a family background that was negative for the disease, the child was diagnosed as a hemophiliac when he was 6 months of age after the discovery of a hematoma on his right forearm This was followed, 2 months later, by severe bleeding into the back, which required a hospital admission. A hemorrhage into the right knee, the first articular involvement, occurred at the age of 3 after an injury, and led to the development of a flexion contracture. Transfusions stopped further bleeding into the joint but traction failed to correct the flexion contracture.

At the time of his first orthopaedic examination on March 22, 1946, the child was confined to bed because of his right knee, but appeared in otherwise good general condition. Extension at the knee was limited to 150 degrees but flexion was free The joint exhibited considerable crepitation on passive motion, and the musculature of the entire right leg was atrophic. Roentgen examination showed a typical hemophilic arthropathy with complete destruction of articular surfaces and conspicuous involvement of the patella. The left knee was entirely normal

For the next 3½ months, the patient was treated on an ambulatory basis for the flexion contracture of the right knee. In future cases, most orthopaedic corrective treatment was to be handled in the hospital but this patient's parents had specifically requested that the boy be treated at home and had rented an apartment near the hospital to facilitate the arrangement Treatment was begun on March 28, 1946 with application of a carefully padded hinged cast to the entire right leg The following day, mild corrective forces in the form of six elastics were added, and in less than 2 weeks the contracture had yielded to the point where extension to 170 degrees had been obtained The hinged cast was then converted into a maintenance cast and fitted with a walking stirrup, enabling the patient to become ambulatory By April 25, he was walking very well and lateral x-rays showed extension at the knee to 165 degrees As there was definite evidence of posterior subluxation of the tibia in relation to the long axis of the femur, and bony contact of the anterior articular surfaces of femoral and tibial condyles, further extension of the knee was excluded because of the risk to the soft femoral condyles from the harder margin of the tibial plateau The leg was then splinted until a plaster of Paris model could be taken for a Hessing-type brace that was jointless at the knee but designed with an ankle joint This procedure, together with the subsequent application of a new walking cast, was carried out under a general inhalation anaesthesia as the patient had become quite apprehensive When this molded leather Hessing-type brace, the first of its kind, was delivered, it included a light-weight foot section constructed with only one medial upright bar connecting the lower-leg section to the simple foot plate

By July 10, the patient was walking well and had started to swim without the appliance An examination at that time showed no evidence of swelling or contracture at the knee A month later, however, a tendency toward flexion had developed as the patient had been sleeping without his brace, contrary to instructions. As a result, x-rays taken on October 3, showed further destruction of the articular surface of the joint with definite decrease in articular space

During this period, the child had required overnight hospitalization and blood transfusions for an extensive hemorrhage into the abdominal wall On the orthopaedic level, an attempt was made to correct the marked valgus position of the feet by providing the patient with a new foot section for the Hessing-type brace and with a Whitman-type plate for the left foot

Treatment from the beginning of October through the end of the year was retarded by a series of hemorrhages into arms, left knee, left ankle and the right leg The patient's clotting time was very prolonged and he required several transfusions To improve the markedly atrophic musculature of the entire leg, particularly the quadriceps, the child's parents were instructed in the use of an electric machine for sinusoidal stimulation This treatment, however, had to be discontinued within a short time as it appeared that electric stimulation of the musculature was responsible for recurrent hemorrhages into soft tissues On January 14, 1947, a new Hessing-type brace with lock joint for the right knee was delivered and immediately provided considerable improvement in the patient's gait His range of physical activities was increased and horseback riding was

specifically permitted in the hope that exercise in the stirrup might increase flexion at the right knee.

Continuing to alternate between activity and disability, the boy was again incapacitated a week later in a fight with another child when he suffered a hematoma at the right knee so extensive that he was forced to discard his orthopaedic appliance As a result, 25 degrees of extension at the knee were lost and the joint exhibited swelling and local heat upon examination on January 30 To correct the deformity, the Hessing-type brace was fitted with crossed elastics which succeeded in straightening the knee sufficiently within two days to permit locking of the knee joint of the brace. With the knee held fully extended, the patient began to walk quite well. On April 10, 1947, however, the ring lock at the knee joint of the brace suddenly became loose when the patient fell out of bed and the knee was forced into flexion By mid-May, however, he was once again playing and running without restrictions. On May 22, another attempt was made to stabilize the knee by substituting an artificial quadriceps for the ring lock of the brace The patient also received a celluloid night splint for the right leg to hold the knee at AGE 170 degrees and the ankle at AGE 90 degrees. Upon examination on July 23, the boy was extremely active and, at the same time, able to maintain extension at the knee to 170 degrees. As the orthopaedic appliance for the leg had become too small, a third Hessing-type brace was made to a plaster of Paris model and delivered on November 11, 1947.

During these 18 months, the child's neurotic tendencies, which were to complicate treatment throughout childhood and early adolescence, had already come into the foreground. As he was an only child and afflicted with a severe degree of hemophilia, he was often isolated from other youngsters and hence largely dependent upon his young parents for companionship and education as well as for physical care. It was quite natural that his parents would try to adjust the child's physical activity to his hemophilic condition It might be said that when the child was not completely immobilized with severe articular or internal hemorrhages, he was encouraged to engage in the most dramatic activities that careful planning and money could provide, including horseback riding, bicycling, driving a tractor, fishing, and living on a boat. The fact that the patient's mental behavior reflected these drastic vicissitudes might be another way of saying that the boy was reacting normally to an abnormal living situation where the feast or famine pattern of activities could only have produced behavior problems

Following recurrent hemorrhages into the right ankle at the end of April 1948, a new Hessing-type brace was constructed At the time it was delivered, the leg was held fully extended, and the knee and ankle joints were completely asymptomatic

On July 18, 1948, the boy suffered a severe hemorrhage into the left calf After a week's hospitalization for hematological treatment, he developed a flexion contracture at the left knee and was unable to walk. Presenting himself for orthopaedic treatment on August 20, he was still totally disabled, having suffered another hemorrhage into the left thigh. Clinical examination revealed a tendency toward flexion at the left knee although it could be fully extended, and roentgen

examination indicated that all osseous structures had remained normal Increased tissue density in the popliteal region suggested residual fluid from the recent hemorrhage. Films of the right knee showed good alignment of femur and tibia although there was a possibility of bony contact between tibia and femur in the center of the knee The skeletal structure of the right leg was much thinner than that of the left

Temporarily, the left leg was splinted with the hinged cast made originally for the right leg As this proved unsuccessful, weighting of the knee was advised

By June 1949, the patient had grown considerably, appeared much more cooperative, and had been leading an active life An orthopaedic shoe was substituted for the Whitman-type plate for the left foot Examined again in the fall of 1949, he reported that he had had an excellent summer As the right knee had remained painless and almost completely extended, the old Hessing-type brace was considered unnecessary. Instead, the patient was provided with a double-bar brace for the right leg with a medial bar extended down to the ankle and attached to a Whitman-type foot plate

Although there had been several hemorrhages into the left ankle, motion at the joint was only slightly restricted, and protection by an elastic bandage was deemed sufficient

When next seen on December 13, 1950, the patient was 9 years old, very wild and difficult to manage He walked with the entire right leg stiff and with a limp that had not been noted before. The right knee was almost completely rigid in extension of 160 degrees and the musculature of the leg was quite atrophic

Roentgen examination shortly thereafter showed continuing underdevelopment of the skeleton of the right leg with considerable decalcification of osseous structures of the knee There was complete destruction of articular cartilage throughout the knee and articular surfaces of femoral and tibial condyles were quite irregular and incongruent although some articular space was preserved between the condyles, and general axis alignment in the sagittal plane was good Lateral exposures showed an angle of 165 degrees and the possibility of minimal anteroposterior motion There was no tendency toward bony ankylosis as previously suggested by films inspected on August 20, 1948

The left knee remained free of involvement with hemophilic arthropathy, and there was no indication of the disease at either ankle

The examination was followed by two hospital admissions in rapid succession for large hemorrhages around the left knee requiring several transfusions A series of x-ray films showed no pathologic changes of the bone but the presence of fluid in the joint was well visualized

By May 17, 1951, the musculature of the right leg had gained in volume and the old *Hessing-type brace had become too small For this reason,* and also because motion at the knee was possible from AGE 170 degrees to AGF 155 degrees, the patient required a new appliance for his right leg A fifth leg brace was forthwith constructed, with Lofstrand spring joints at the knee for the first time Henceforth, this appliance was to be known as a hemophilia Hessing brace

For 6 weeks prior to his next orthopaedic consultation on October 1, 1951, the patient had been bedridden or wheelchair ambulatory with swelling at the left knee and right ankle. He had required the services of a visiting teacher, and his psychological problems were again in the foreground as he had been isolated from other children for a long time. Roentgen examination revealed the first signs of early hemophilic arthropathy at the right ankle caused by incongruent axes of anatomical and brace ankle joints There was also an obvious lack of skeletal development in comparison with the normal left ankle and moderate decalcification at the astragalus A plaster of Paris model was immediately taken of the right lower-leg for a new brace section with a properly placed ankle joint At the same time, the left knee required attention because of recurrent hemorrhages and ballottement of the patella Orthopaedic treatment was initiated with complete immobilization in a protective plaster of Paris cylinder. By the middle of October, the patient's gait was considerably benefited by a Hewitt elastic knee brace with posterior hinge and one-piece tongue for the left leg, and a reconstructed hemophilia Hessing brace for the right leg with a new foot section consisting of a molded leather sandal with a plain foot plate, stirrup and milled ankle joint

Toward the end of 1951, orthopaedic treatment was punctuated by several major bleeding episodes affecting ankles, left knee, left arm and face, requiring three hospital admissions for hematological treatment. At one point, the Lofstrand spring joints of the right hemophilia Hessing brace were replaced by a solid jointless connection holding the knee at an angle of 170 degrees as even minimal motion was poorly tolerated. The patient also received a jointless molded leather-steel brace for the same leg to be worn as a night splint or used during the day when the ankle did not need special support.

With a recurrence of pain and swelling at the right ankle, roentgen examination again revealed a discrepancy between anatomical and brace ankle joints, responsible for these symptoms as well as other locomotion difficulties The brace was accordingly altered to provide the proper alignment. Films of the right ankle showed that the distal epiphysis of the tibia had not grown as much on the fibular side as on its medial aspect This caused a rather marked tilt of the mortise and the astragalus into valgus position. The capsule of the joint showed increased thickening at two points.

During this period, a plaster of Paris model was taken for a jointless molded leather lower-leg brace for the left leg with attached Whitman-type foot plate. A protective cast was applied to the left leg until the appliance could be delivered

Spending the winter of 1952 in Florida, the patient was largely wheelchair ambulatory because of recurrent hemorrhages into the left knee and thigh. On June 12, 1952, at the next orthopaedic examination, the patient's legs appeared greatly improved, with the right knee joint extended to 170 degrees and capable of a few degrees of active flexion He was able to swim every day and to walk around the pool without his braces Another Whitman-type plate was made at this time for the left foot.

Toward the end of 1952 and the beginning of 1953, the patient suffered a

series of hemorrhages into the left knee which was no longer properly supported by the Hewitt elastic knee brace. To further complicate the case, roentgen examination on November 13, 1952 revealed first signs of involvement of the left ankle with an early stage of hemophilic arthropathy The orthopaedic plan to protect the entire leg by means of a plaster of Paris cast was opposed by the attending hematologist who finally agreed to immobilization of the left lower extremity on a plaster of Paris splint. As soon as the elastic motion-permitting Hewitt knee brace was reapplied, hemorrhages into the knee recurred, and another hospital admission was required for further immobilization of the joint and administration of plasma This time, the patient was explicitly cautioned to refrain from weight-bearing until an appliance could be constructed for the left leg corresponding in every detail to the hemophilia Hessing brace with Lofstrand spring joints at the knee that had just been manufactured for the right leg The brace for the left leg was finally delivered together with a small fiberglass cylinder to be worn at the beach and while swimming in Florida during the winter

While in the South, the child suffered a series of hemorrhages into the right groin and developed a mild flexion contracture causing a back disability Shortly after his return, he hemorrhaged severely into the left knee while sleeping with an elastic knee brace instead of the hemophilia Hessing brace Examination during the subsequent hospital admission for plasma transfusions revealed a marked degree of lateral instability caused by distension of the capsule of the left knee joint

When examined in June 1953 the boy had grown tremendously and his musculature showed excellent development Roentgen examination indicated for the first time definite evidence of involvement of the femoral condyles of the left knee joint with early hemophilic arthropathy The previously noted lateral instability with the left knee extended was still present

The patient arrived wheelchair ambulatory for a further examination two months later The hemophilia Hessing brace for the left leg had been discarded following recurrent hemorrhages into the knee and the joint was supported only by an elastic bandage While the parents preferred to keep the child wheelchair ambulatory as long as the tendency toward hemorrhages into the joint persisted, this was considered undesirable from an orthopaedic point of view as development of all structures of the extremity might be retarded if it remained inactive for too long a period

The patient claimed that he could not maintain his balance with two hemophilia Hessing braces and was unable to tolerate the action of the Lofstrand spring joint on the right side Concentrating, therefore, on the important problem of left knee instability, it was decided to continue with the left hemophilia Hessing brace, and the appliance was fitted with an ischial seat to transmit weight-bearing from the tuber ischii to the ground The right hemophilia Hessing brace, however, was replaced by a Hewitt elastic knee brace This was practicable as the right knee had remained asymptomatic ever since the first Hessing-type leg brace had been applied more than 7 years earlier

Despite these measures, the child was hospitalized again on September 22,

1953 for new and painful hemorrhages into the left knee and thigh For the first time, the patient did not respond to plasma transfusions as he had developed an anticoagulant which interfered with the antihemophilic action of the plasma

For four months after the patient was discharged from the hospital in a wheelchair on September 26, 1953, treatment was concentrated primarily upon the left leg After a cast was applied holding the leg completely extended, swelling of the knee and thigh subsided and ballottement of the patella disappeared, proving once again that complete immobilization in a well padded plaster of Paris cast was the most effective method of preventing additional hemorrhages and obtaining absorption of fluid within the joint Following application of a walking cast, the reconstructed hemophilia Hessing brace was reapplied and the patient became ambulatory.

An attempt to use the right hemophilia Hessing brace without its foot section but with a single Lange-type fiberglass plate for the right foot failed because there was insufficient stability at the right ankle which was held in marked valgus position due to difference in the development of the medial and lateral aspect of the distal tibial epiphysis Instead, a new hemophilia Hessing brace that was lighter in weight and less bulky was constructed with milled ankle joint but without knee joint.

Examined again on April 14, 1954 after his return from Florida, the patient stated that he had enjoyed a good winter with only two minor hemorrhages into the left ankle. He walked comfortably and all indications of lateral instability had disappeared The left knee was fully extended and the right knee maintained extension at 170 degrees On June 3, the patient reported that he had walked occasionally without his braces and had had no further hemorrhages or pain at either knee Major appliances were completely discarded on June 7, but the right knee was fitted with a new Hewitt elastic knee brace to be worn continuously because of x-ray evidence of involvement with marked hemophilic arthropathy. Despite instructions, the patient stated 2 months later that he had not used any orthopaedic appliance during the summer. The condition of both legs was satisfactory and the boy was planning to attend school

For more than 2 years between May 1954 and July 1956, the patient had not required orthopaedic treatment. Apart from a large hemorrhage into the left knee joint in September 1955, necessitating a flight from Florida to New York for transfusions and observation, there had been no pain or disability originating from the knee joints and the patient had worn no orthopaedic appliances For recurrent hemorrhages into and around the left ankle with pain and disability of short duration, he again required plasma transfusions but no further treatment

In the fall of 1955, he had been hospitalized in critical condition for three months with a perforated appendix, peritonitis, an intestinal obstruction and hemorrhages into the abdomen. Eventual recovery was made possible only through the use of antibiotics and continuous administration of whole blood and plasma transfusions

When examined on July 10, 1956 after a 2-year interval, the boy, then 14

years old, was bedridden with an acute locking of the right knee at AGE 155 degrees In the interim, he had grown considerably, and appeared much more mature in every respect. According to the patient, the right knee became quite stiff when exposed to dampness or cold Examination suggested locking from part of a torn internal semilunar cartilage There was no evidence of hemorrhage

Although the patient had recurrent hemorrhages into the left ankle, he refused to wear adequate shoes. The left knee joint had remained asymptomatic

With immobilization of the right knee on a large posterior splint, pain was minimized during the day but toward evening became so severe at times that the patient required Codeine and Aspirin and was unable to sleep Relief was finally provided by application of an extra-long foam rubber pad on the dorsum of the leg Shortly thereafter, the joint became asymptomatic and by July 20, 1956 had returned to its pre-locking condition, in extension of 170 degrees A maintenance cast was applied and split in the anterior midline to permit easy removal at home Remaining normal for a week in spite of greatly increased activities, the leg was fitted with a 20-inch Hewitt elastic knee brace with double steel bars in the back and one-piece foam rubber padded tongue

Complete roentgen examination of knees and ankles on July 17, 1956 showed extensive hemophilic involvement of the right as well as the left knee While the right knee presented evidence of a permanent "burned out" condition, the left knee had also suffered extensive damage Articular space at both knees, however, was surprisingly well preserved despite considerable cartilage destruction

Ankle joints also showed involvement with hemophilic arthropathy, more marked on the left side The medial epiphysis at the distal end of the tibia had grown faster than the lateral causing the usual valgus deformity, again more marked on the left side

Comparing these films with a series taken 3 years earlier, there appeared to be satisfactory development of osseous structures of the lower extremities Shafts of femur, tibia and fibula were of normal size and calcium density with a strong cortex Pathology was limited to the epiphyses and articular surfaces The right knee, which had experienced no hemorrhages in 10 years, had developed more fully than anticipated It was particularly fortunate that the shafts of femur and tibia had grown so strong and that there was no significant degree of decalcification It was apparent that the right knee would remain smaller than the left as epiphyseal lines were closing prematurely with the undersized patella exhibiting a typical hemophilic deformity The apophysis at the tibial tubercle was completely fused on the right side while it was still open on the left Considering the "burned out" condition of the right knee, support by means of a Hewitt elastic knee brace was considered sufficient

The left knee and ankle, however, required stronger orthopaedic protection Even if there were to be no more bleeding into these joints, articular surfaces exhibited so much deformity, incongruence and irregularity that unprotected weight-bearing would most certainly lead to synovitis and arthritis and cause great pain in later life Although further development of secondary degenerative

joint disease at the left knee and ankle might not be prevented, it could be somewhat contained if *weight-bearing is permitted only upon fully protected joints*, at least until the end of the growth process. With the patient's reluctant acceptance of the plan for permanent bracing of his left leg, a hemophilia Hessing brace with milled knee and ankle joints was made to a plaster of Paris model, and delivered on July 31, 1956.

Summary: Reviewing the 10-year period of observation and treatment of a severe hemophilia sufferer, 12 protective and corrective plaster of Paris casts and splints were applied, and 16 plaster of Paris models were made for the manufacture of 10 Hessing-type and hemophilia Hessing braces, 1 molded leather lower-leg brace, 4 foot plates and 1 fiberglass appliance Treatment also included the use of 6 smaller orthopaedic appliances that were not made to plaster of Paris molds The patient required 11 hospital admissions for extremity hemorrhages as well as many additional hospitalizations and countless transfusions for other hemophilic problems

The original objective of orthopaedic treatment was the correction of a flexion contracture of the right knee joint After successful treatment of this deformity, almost complete extension of the knee was maintained without a recurrence of pain or bleeding into the joint even during periods when orthopaedic appliances were discarded

Treatment was not successful, however, in preventing the development of hemophilic arthropathy of the left knee and, to a lesser degree, of the ankles with their marked valgus deformity. There were several reasons for this failure First of all, orthopaedic rehabilitation of hemophilic arthropathies had not developed to its present standard during the first 5 years of the boy's treatment Second, quick-frozen antihemophilic plasma was not available until more recently Also, in this case, the parents sought to achieve the most satisfactory balance between psychological needs for activity and physical restrictions, and tended to encourage activities that entailed considerable risk for the patient Furthermore, the earlier braces were not yet as well adapted to the special requirements of hemophiliacs, and there was obviously some inconsistency of treatment From today's orthopaedic point of view, correction of the flexion contracture of the boy's right knee by wedge or Quengel cast should have been followed by uninterrupted immobilization in a well-fitting hemophilia Hessing brace until osseous structures were fully developed, epiphyses closed and the musculature in condition to control whatever motion was still present at the knee Whenever hemorrhages threatened any of the other articulations, such as left knee and ankle joints, immediate plaster of Paris immobilization should have been instituted in addition to administration of plasma to stop the bleeding This would have prevented joint destruction, particularly if bracing of the extremity had followed plaster of Paris treatment The importance of orthopaedic appliances for prevention of joint involvement in hemophilia was not clearly recognized until quite recently, while their function in rehabilitation and maintenance of correction obtained by plaster of Paris treatment was acknowledged from the very beginning This case history clearly demonstrates the point that it took a

long time before the responsibility for prophylaxis and treatment was shared equally by hematologist and orthopaedic surgeon In the earlier years, too much reliance was placed on transfusions alone and it was difficult to convince hematologists of the necessity and feasibility of application of protective plaster of Paris casts during the acute phase of extremity hemorrhages

Treatment was rendered more difficult in this case by the unfortunate local division of treatment between one hospital for hematology and another for orthopaedic treatment. The hemophilic involvement of joints other than the originally affected right knee might well have been prevented if the orthopaedic surgeon had been in a position to offer treatment from the very first sign of a hemorrhage instead of being called after the hemorrhage had been halted by bed rest, refrigeration and transfusions

Despite these drawbacks, the patient's physical condition was satisfactory, with almost normal development of the skeleton of the lower extremities except for the hemophilic joints Because the right knee joint had been treated when the patient was 4 years old, and unprotected weight-bearing on the right leg was not permitted for 7 years thereafter, the development of the genu valgum by overgrowth of the medial femoral condyle, seen in almost every other hemophilia patient, was prevented Periodic roentgen examination of the right knee joint has given considerable insight into the structural changes which produce the characteristic x-ray appearance of a hemophilic arthropathy of the knee joint if the disease has occurred during childhood or adolescence

Perhaps more than anything else, this case history demonstrates that complete plaster of Paris immobilization of an extremity during an acute phase of bleeding into joints or soft tissues will promptly eliminate pain and prevent development of contractures and deformities

Case #7185 (See Figures 2 and 18)

Arnold M, an adopted child with no known hemophilia background, had shown hemophilic symptoms since infancy and developed hemophilic arthropathies of the right knee and left elbow as well as a moderate degree of bilateral spastic equinus during early childhood At his first orthopaedic examination, the boy, then 7 years of age, stood and walked chiefly on metatarsal heads with a moderate bilateral adductor spasm causing a scissors' gait Although both knee joints were quite swollen, only the right showed signs of hemophilic arthropathy Ankles exhibited muscular rigidity The musculature of the left arm was atrophic and the left elbow was deformed and showed restriction of motion At no time had the child received therapy for any of his orthopaedic problems

Examination also revealed an unusually small skull, suggestive of premature ossification of cranial sutures rather than a birth injury In addition, a marked strabismus was noted which could not be operated upon because of hemophilia.

X-ray films of the right knee indicated increased soft tissue density without significant bone or cartilage involvement The patella and epiphysis of tibia and femur were larger than on the left side demonstrating the over-development frequently seen in hemophilic arthropathies Skeleton of feet and ankles was poorly

developed but otherwise within physiological limits Films of the left elbow revealed quite marked deformity of trochlea of humerus and of articular surface at proximal end of ulna The epiphysis of the head of the radius was fairly large and the lower end of the humerus was also enlarged

During a 21-day hospital admission from September 9 to September 30, 1954 for orthopaedic correction of hemophilic arthropathies and bilateral spastic equinus, a heavily-padded cast was applied to the right leg from toes to below the knee holding foot in best possible dorsiflexion of 100 degrees while the knee was flexed to 90 degrees. This procedure was intended to test the response of the spastic right leg to rigid immobilization and was well tolerated without producing increased spasticity Patient was able to walk with leg in the cast although he was still holding feet in equinus position. This was followed by application of a lighter cast to the right leg in two sections. To correct equinus position, foot was held at dorsiflexion of 100 degrees and cast was applied from toes to below knee When this section became rigid, the knee was fully extended and the cast completed to below the groin.

During this period, x-rays of the skull showed moderate increase in digital markings of cranial vault, and evidence of early closure.

After termination of this first phase of plaster of Paris treatment, patient received three orthopaedic appliances These consisted of a fiberglass brace to support and protect the left elbow, with an elbow joint permitting immobilization of the elbow at various degrees of flexion by means of a locking device, a hemophilia Hessing brace for the right leg with Lofstrand spring joints at the knee, milled ankle joint with stop to prevent plantar flexion and with crossed elastics to increase dorsiflexion, and a fiberglass Lange-type foot plate for the left spastic foot to which a posterior drop foot spring with calf band was attached Despite patient's spasticity, appliances for the lower extremities were well tolerated and served to improve stance and locomotion. Patient no longer walked with both feet in equinus position but with heels well on the ground

Eight months later, patient was again admitted to the hospital to be fitted for new orthopaedic appliances for his legs. Examination revealed no new signs of hemophilic arthropathies or hemorrhages into soft tissue. Spasticity of lower extremities was less marked than previously noted. Roentgen examination showed that osseous structures of knee joints had grown since September 1954 and epiphyses had developed normally. The only change in structure of the right knee since that time was the considerable thinning of the proximal third of the shaft of the fibula.

During this admission, the patient received a new hemophilia Hessing brace for his right leg together with a similar appliance for the left leg as the single fiberglass plate was no longer sufficient. With these new appliances, there was immediate improvement in his gait

A year later, the boy was re-admitted for the third time following acute hemorrhages into left knee and left elbow Parents of the patient stated that with his legs braced the boy had been able to walk much better than anticipated until a few weeks prior to this hospital admission when the braces appeared too small

and too tight and a hemorrhage into the left knee had occurred Upon examination, the knee appeared greatly enlarged but not very painful During this 10-day hospitalization, plaster of Paris models were taken of both legs and the patient received two new hemophilia Hessing braces with elastic twister straps to be used to counteract the tendency toward internal rotation of legs from spastic equinus Appliances were well tolerated and, at that point, general rehabilitation was better than could have been expected in view of the boy's multiple handicaps

Case #7207 (See Figure 17)

When hemophilia is not spontaneous in origin, it is passed genetically from mother to child The female carrier is supposedly without symptoms of the disease The case of Ernest M was the only one in the series in which a mother reported a bleeding tendency that appeared after an early tonsillectomy and adenoidectomy, and also during childbirth Aside from this, there was no family history of hemophilia

Although the boy hemorrhaged severely at circumcision, he was not diagnosed as a hemophiliac until he was 1 year old. Since then, he suffered hemorrhages into elbows, ankles and knees as well as gastro-intestinal bleeding, hematuria, chest hemorrhages and gum bleeding During his early childhood, he was hospitalized on numerous occasions, received twenty-nine blood transfusions, and was given an orthopaedic appliance for his left leg at 4 years of age

At his first orthopaedic consultation, he was 7 years old, crutch ambulatory and in poor health. Clinical and roentgen examination revealed severe hemophilic arthropathy at the left knee and a flexion deformity of 90 degrees with some posterior subluxation of the tibia in relation to the femur The right knee showed swelling and local heat due to a recent hemorrhage but no restriction of extension Osseous structures were underdeveloped but there was no articular destruction

The left foot and ankle were improperly developed with considerable decalcification of all osseous structures but no evidence of hemophilic arthropathy The left elbow was inflamed with extension possible to only 125 degrees X-ray films indicated hemophilic arthropathy with hemarthros and showed a large juxta-articular cyst in the olecranon, slight involvement of osseous structures of the capitellum, and quite marked soft tissue swelling around the joint

During a 53-day hospitalization, a maintenance cast was applied to the entire right leg, with the knee fully extended, for complete immobilization during the acute hemarthros When the cast was removed, the knee was asymptomatic and a plaster of Paris model was taken for a hemophilia Hessing brace with Lofstrand spring joints at the knee At the same time, a Quengel cast hip spica with subluxation hinges was applied to the left leg After correction of extension from 110 to 155 degrees was obtained, a model was also made for a left hemophilia Hessing brace with Lofstrand spring joints at the knee Until the new appliances could be delivered, the lower extremities were protected by a strong splint on the left side and a maintenance cast on the right side

As the left elbow had become painless with extension limited to 140 degrees, a model was made of the arm from above the wrist to the deltoid region for a

fiberglass protective splint At the end of this hospital admission, which had been complicated by gastro-intestinal bleeding, all appliances were delivered and well tolerated

Examined 18 months later, the patient had grown considerably, was attending school, and showed additional extension at the left knee to 165 degrees. Although there had been no further bleeding into knee joints, roentgen examination revealed very slight involvement with hemophilic arthropathy at the right knee The right elbow had remained normal despite a recent hemorrhage into the soft tissues, while the left elbow exhibited slight but not significant progression of deformity from hemophilic involvement.

Models were made of both legs for new lower-leg sections for the hemophilia Hessing braces which were reconstructed and delivered 5 days later, together with an elastic attachment for further extension at the left knee.

A year later, the patient was hospitalized for 10 days while new hemophilia Hessing braces and a fiberglass brace for the left elbow were manufactured to plaster of Paris models When discharged from the hospital on June 28, 1956, the patient, free of bleeding at the knees since he had first worn hemophilia Hessing braces, was considered completely rehabilitated with his new appliances

Case #7301

Philip C, age 7, with a hemophiliac brother and two sisters, was diagnosed as a hemophiliac during infancy and thereafter afflicted with recurrent bleeding Hemorrhages into joints were quickly absorbed, leaving no deformity or disability, until September 1954 when a severe hemorrhage into the right knee did not respond to plasma transfusions Treatment of the joint was complicated by a serious hemorrhage into the oral cavity that interfered with swallowing and breathing. During a hospital admission for this episode, numerous transfusions had little influence on the hematoma but bleeding gradually subsided, probably due to pressure The right knee joint, neglected during this period, remained enlarged with restriction of extension.

When the boy appeared for his first orthopaedic examination on November 15, 1954, he was cane ambulatory, walking with a right-sided limp, holding the knee and hip in flexion and the foot in equinus position. The musculature of the entire right lower extremity was in good condition, showing very little atrophy The knee joint was enlarged, partly due to hemarthros, partly to enlargement of femoral condyles and showed extension to 160 degrees and local heat. There was also discoloration of the skin in the region of the right iliac crest

Roentgen examination revealed minimal involvement of the articular margin of medial and lateral femoral condyles as well as increased density of soft tissues around the knee. A small cystic area on the posteromedial aspect of the femoral shaft and widening of femoral condyles were additional signs of early involvement with hemophilic arthropathy

The patient was accordingly given a Hewitt elastic brace for the right knee

and by the end of November 1951, the joint had become almost completely extended, and the patient walked very well

Although he had been completely free of hemophilic arthropathies at the left knee and right ankle, neglect of recurrent hemorrhages into those joints during the interval between orthopaedic examinations, led to permanent damage. While orthopaedic treatment at the first indications of hemarthros could have prevented articular destruction, the child's parents relied exclusively upon temporary bed rest and multiple transfusions Consequently, when the patient appeared for his first orthopaedic examination in eighteen months, he exhibited marked hemophilic arthropathies of left knee and right ankle Maintenance casts were immediately applied to the entire left leg and right lower-leg during an overnight hospitalization Thereafter, as a major appliance was required for the left leg, a hemophilia Hessing brace was made to a plaster of Paris model and delivered on June 28, 1956

Case #6607

By the time Daniel W was 6 years old, he had become a severe hemophilia victim afflicted with recurrent internal bleeding and extremity hemorrhages complicated by a consistently high clotting time As there had been a long history of the disease in the mother's family, diagnosis of hemophilia was established at the birth of the child with the appearance of two cephalohematomas During infancy, he suffered frequent hemorrhages into both legs, gastro-intestinal bleeding as well as hemorrhages following an ear infection and head injuries At the age of 3, he was hospitalized with hemarthros of the left knee which persisted despite transfusions, and eventually produced a difference in length of legs as the left leg grew faster Two years later, an injury to the left knee produced bleeding into articular space and caused a 5-10 degree flexion contracture Also during these early years, the left elbow became stiff, and an injury to the right flank required hospitalization and many blood transfusions

In March 1953, 5 months before his first orthopaedic examination, he suffered hemarthros of the right ankle Treated with an injection of Hyaluronidase and transfusions of antihemophilic plasma, bleeding was stopped and a normal range of motion restored at the ankle

As the father's profession involved repeated change of location for the family, treatment of the child was sporadic and inadequate Orthopaedic treatment, begun in February 1950 for hemarthros of the left knee, consisted of diathermy twice daily for 1 month and then intermittently for several months Corrective shoes, incorporating a compensatory lift on the right side, were then recommended because of inequality in length of legs Physiotherapy was resumed in March 1953 because of marked restriction of motion at the left knee Patient also wore metal splints for protection of knee joints

Since December 1952, the patient had received antihemophilic plasma transfusions at 2- to 4-week intervals which kept bleeding under fair control.

At his first orthopaedic examination on August 27, 1953, the patient was

crutch ambulatory and unable to bear weight on the left leg. The left knee showed a flexion contracture of 80 degrees with marked structural deformity of femoral condyles and patella, and posterior subluxation of tibia. The left elbow also was hemophilic with restriction of extension and flexion

Inspection of original x-ray films of the left knee dated July 15, 1953 revealed considerable irregularities of femoral condyles most marked at medial articular surface There was no definite evidence of juxta-articular cysts Patella showed squaring off at its distal pole. Articular space between patella and femoral condyles was diminished while space between femoral and tibial condyles seemed well preserved

New x-ray films showed considerable progress of hemophilic arthropathy at the left knee Femoral, and to a lesser degree, tibial condyles were blown up and showed more decalcification than they had 6 weeks previously. The patella appeared glued to femoral condyles There was some posterior subluxation of tibia Tunnel view showed marked reduction in width of articular space between femur and tibia. Tunnel view of the right knee showed some structural irregularities at lateral more than at medial femoral condyle which may have represented an early stage of hemophilic arthropathy.

The left elbow exhibited severe hemophilic involvement without generalized decalcification of bones There was increased soft tissue density at the joint. Several small calcific shadows were visualized within the joint apparently between articular surfaces on the ulnar side. While condyles of the humerus showed no bone destruction, there was considerable overgrowth in comparison with the right elbow, and the articular surface of the ulna showed marked deformity and enlargement at the expense of the olecranon. The proximal epiphysis of the radius was very large and dense

For orthopaedic correction of the 80 degree flexion contracture of the left knee, the patient required carefully planned treatment extended over a sufficiently long period to avoid fatigue which might increase his already high clotting time During a first hospital admission of 18 days from August 31 to September 18, 1953, a Quengel cast hip spica was applied to the left leg from umbilicus to toes with knee held in flexion of 80 degrees and ankle at 90 degrees. No attempt was made at the time to correct the valgus tendency at the knee. After the cast was removed, with extension of the knee having reached 140 degrees, a maintenance cast was applied to enable all soft tissue structures of the left knee to adjust to the new position After sufficient time was allowed for this adjustment, the maintenance cast was replaced by a second corrective cast which was prepared for wedging at the knee. Following a single wedging of the cast, the patient was discharged wheelchair ambulatory with completion of this first phase of corrective treatment to be handled on an ambulatory basis. After another wedging, the cast was replaced by a maintenance cylinder that permitted crutch ambulation. The patient also began to attend school. As extension of the knee, however, remained limited to 140 degrees, a second wedge cast was applied

During a hospital admission from October 26 to November 3, 1953, scheduled primarily for extraction of an abscessed tooth, x-rays of the right foot and ankle

were taken and showed marked soft tissue swelling around the ankle resulting from a hemorrhage into the joint. Osseous structures appeared rather poorly developed but otherwise normal. Plasma transfusions were administered and a maintenance cast was applied to the foot and ankle until the inflammation had subsided.

Also during this period, hemorrhages into the right eye seemed to be interfering with the patient's eyesight Ophthalmological examination revealed a scar in one eye which may have been due to an old hemorrhage but which did not impair vision too much The other eye was normal

When the wedge cast which had remained in place on the left leg since mid-October was removed on December 1, 1953, extension at the knee had reached 150 degrees and a plaster of Paris model was finally taken of the leg for a hemophilia Hessing brace Thereafter, a well-padded maintenance cast was applied to the entire extremity permitting some weight-bearing and crutch ambulation After the appliance was delivered, Lofstrand spring joints at the knee were temporarily locked at 155 degrees until the leg could become accustomed to the brace and the knee joint could tolerate corrective forces introduced by springs. A knee cap was also added for protection A week later, the patient walked well with the left knee rigid in flexion of 150 degrees Screws locking the action of the Lofstrand joints were removed and the springs were well tolerated

A further examination on January 28, 1954, prior to the boy's departure for California, showed considerable improvement at the left knee which was extended to 160 degrees, and the patient was walking comfortably

When examined upon the patient's return to New York City 8 months later, the left knee could be extended to 170 degrees The hemophilia Hessing brace, however, had become too small since it had been made almost a year previously and had also suffered from several attempts at adjustment while the child was living on the West Coast A second hemophilia Hessing brace was therefore constructed to a plaster of Paris model.

As a series of painful hemorrhages into the soft tissues of the right hand had caused marked restriction of motion at the wrist, immediate splinting of the right forearm, from the proximal phalanges to below the elbow, was required

A protective fiberglass splint was also made for the left arm as there had been recurrent hemorrhages into the elbow

On November 12, 1954, the patient was again admitted to the hospital for an acute hemorrhage into the left elbow which responded promptly to plasma transfusion Protected by an elastic bandage, the joint appeared to be asymtomatic at the end of a 4-day admission

A week later, however, the boy had to be readmitted for recurrent hemorrhages into the left elbow, an acute hemorrhage into the right calf and a slight hemorrhage into the upper lid of the right eye. After a transfusion with antihemophilic plasma and application of ice, the hemorrhage into the right calf subsided but swelling had caused a tendency toward equinus position of the right foot, and the right knee could not be completely extended. An elastic bandage was therefore applied from toes to mid-thigh and the patient was instructed to continue

on bed rest until all swelling had disappeared and the right knee could be completely extended. Also during this 4-day admission, the simple fiberglass brace for the left elbow was reconstructed and the joint became painless and freely movable

Examination at the end of June 1955, before the patient was scheduled to leave for a year in California, showed that the left knee had remained asymptomatic and almost completely extended despite clinical and roentgen evidence of severe hemophilic arthropathy. New x-rays, however, indicated that there had been no progression of bone or cartilage destruction since November 1954. Full extension of the left knee was obtained after a new lower-leg section was constructed for the hemophilia Hessing brace. The patient was also given a new fiberglass brace for continuing protection of the left arm until his next examination which was scheduled 1 year later

Case #7762 (See Figure 33)

Frederick J, age 18, with a hemophilic history complicated by focal epilepsy and a freak accident, demonstrated a remarkable degree of rehabilitation following orthopaedic treatment of a fractured hemophilic knee joint

With no evidence of the disease in his family background, this patient experienced his first hemorrhage at circumcision and required three transfusions at that time At 3 months of age, a contusion led to a hematoma on the sole of the foot As soon as the child started to walk, he fell frequently and was never without large hematomas on his forehead. Age 2 marked the appearance of Frederick's first articular hemorrhage, involving the right elbow. A year later, his left arm was caught in the wringer of a washing machine and severely injured, with massive bleeding, although no bones were damaged Despite 2 weeks of hospitalization and continuous bed rest at home for many weeks thereafter, the patient developed Volkmann's ischemia with claw hand. It was during this prolonged period of inactivity that the patient first exhibited symptoms of chorea minor. These disappeared, however, after he was no longer bedridden.

At 9 years of age, recurrent hemorrhages into the left knee joint produced a flexion deformity After unsuccessful treatment by means of traction, the contracture was largely corrected by manipulation under anaesthesia followed by 6 weeks' immobilization of the leg in a cast. An orthopaedic appliance was worn on the left side for 3 years and then discarded with no resultant discomfort or disability despite a slight residual deformity at the knee.

Following 6 months of dizzy spells, the boy suffered his first epileptic seizure with convulsions in June 1953 An electroencephalogram confirmed the diagnosis and Dilantin® sodium was prescribed Nevertheless the seizures continued, with the most recent occurring just prior to his first orthopaedic examination in August 1955 The etiology of this condition might be traced to a fall suffered when the patient was 5 years of age An injury to the back of his skull may have caused a small subdural hematoma leaving scars responsible for the recent epileptic seizures At the time, there were no signs of bleeding or concussion, but a week after the fall, the patient was found unconscious. During a week's hospitalization, a hematoma was located in the left occipital region.

In June 1955, the patient suffered an attack of severe pain in the lower back After treatment with radiant heat, injections and Meticorten® tablets, the condition subsided but recurred in even more severe form a week later During a 5-day hospitalization, the spine was x-rayed and the condition diagnosed as arthritis

Recurrent hemorrhages into the right knee, starting when the patient was 11 years of age, culminated in an acute inflammation of the joint following an epileptic convulsion on August 6, 1955 Regaining consciousness after the seizure, the patient experienced such excrutiating pain in the joint that he was immediately sent to a hospital where he received morphine, blood transfusions, and several injections of Hyaluronidase and Hydrocortone into the right knee In this condition, the patient was discharged from the hospital with the inflamed knee supported only by a posterior splint

At the time of our first examination on August 26, 1955, the patient was wheelchair ambulatory and in poor general condition with very severe pain originating from the right knee which was tremendously swollen, tender and hot Pending a complete examination, he was hospitalized primarily for treatment of an explosion-type fracture of the right femoral condyles The right leg was immediately immobilized on an extra-long flexible knee splint, ice bags were applied to the knee, and plasma and analgesics were administered

A complete orthopaedic examination two days later on August 28, 1955 revealed that the pain at the right knee was subsiding, although the joint was still greatly enlarged without palpatory evidence of an effusion or of much soft tissue swelling The slightest motion caused pain

At the left knee there was very little deformity with a range of active motion from AGE 170 degrees to AGF 90 degrees and a moderate degree of posterior subluxation of the tibia This joint had remained asymptomatic since the patient was 12 years of age

The left forearm and elbow exhibited a moderate degree of Volkmann's contracture without sensory impairment Physical therapy for the past 18 months had brought some improvement in the function of the left hand, and the patient was now able to use it quite well despite stiffness of the wrist and flexion contractures of the second and third fingers due to shortening of the flexor tendons "Automatic transmission" of wrist motion to fingers was well demonstrated The forearm was held in almost complete pronation The musculature showed marked atrophy The appearance of elbow and wrist was due to the ischemia following a massive hemorrhage into the forearm and there was no suggestion of involvement with hemophilic arthropathy

Although the right elbow was asymptomatic, there was evidence of marked involvement with hemophilic arthropathy

Examination of the spine revealed a C-shaped scoliosis convex toward the right side due to a pelvic obliquity with the left leg longer than the right, and with a moderate deformity of the thoracic cage The ribs on the right side appeared more prominent posteriorly

X rays taken the following day showed quite marked generalized decalcification at the right knee joint with increased shadow density of soft tissues, and revealed complete incongruence of the articular surface of the femur and the

tibia due to a recent oblique fracture through the external femoral condyle with rotation and upward displacement of the entire external condyle. The marked widening of the femoral condyles with a central gap into which the tibia protruded *suggested that the tibia was forcefully pushed* between the softened femoral condyles. These films indicated that the severe pain which started after an epileptic convulsion on August 6, 1955 was due to a fracture and not to an acute hemarthros. *Presence of this fracture explained why severe pain in the right* knee persisted after bleeding had stopped.

At the left knee joint there was the characteristic picture of hemophilic arthropathy originating during childhood or adolescence with generalized decalcification, deformity of the patella, thinning of the articular cartilage and some articular and juxta-articular changes There was also some posterior subluxation of the tibia in relation to the femur.

Films of the left upper extremity exhibited mild changes at the elbow joint, most marked at the radiohumeral articulation The deformity at the distal end of the radius and ulna was not suggestive of hemophilic arthropathy.

The right elbow showed a considerable degree of hemophilic arthropathy without soft tissue swelling. Most of the enlargement and deformity existed at the radial head

There was no bone or joint pathology of the spine and no evidence of rheumatoid arthritis. Pelvis and hip joints showed bilateral coxa valga of considerable degree but no bone or joint pathology.

The acute and extremely painful condition of the injured right knee joint made it impossible to carry out the original plan for extensive orthopaedic rehabilitation of the patient's hemophilic arthropathies and corrective treatment of the contracture of the left forearm and elbow.

After a plaster of Paris model for a hemophilia Hessing brace was made of the entire right leg with the knee extended to 145 degrees, the leg was immobilized in a maintenance cylinder As the patient could not tolerate this cast, a posterior flexible knee splint was substituted until another maintenance cast could be applied Correction of the flexion contracture of the knee had to be limited to AGE 150 degrees because of the acute condition of the joint. Further corrective treatment would have to await consolidation and complete healing of the fracture. *After the orthopaedic appliance was fitted and delivered* with the knee joint locked, pain in the knee gradually subsided. Ambulation was started with a small walker and the patient was finally discharged crutch ambulatory *after a month's hospitalization*

During this admission, an electroencephalogram was taken that once again confirmed the diagnosis of epilepsy and revealed an actively discharging epileptogenic focus over the right hemisphere. Films of the skull exhibited no abnormality

Additional x-rays, taken in April 1956 showed marked improvement of the right knee On May 23, 1956, the patient wrote that he had become cane ambulatory *and was able to swim without his brace.*

Case #6268

William M, with a hemophilic family that included two brothers and an uncle, suffered hemophilic involvement of the right knee at the age of 4 This was soon followed by hemophilic arthropathy of left knee and ankles, and for 5 years thereafter the child was totally unable to walk. During this time, he was sporadically admitted to various hospitals but it was not until he was 8 years old that he was subjected to a 13-months' admission for correction of flexion deformities of the knees He was finally discharged from the hospital after both knee joints were fully extended, but he received no orthopaedic appliances whatsoever. It was not surprising that 2 weeks later he fell and suffered fractures of the right lower-leg below the knee and the left lower-leg above the ankle After the fractures healed without significant displacement or deformity, the patient was given double-bar caliper braces with ring locks at the knees He then became crutch ambulatory and his knees remained asymptomatic except for one hemorrhage into the left knee that occurred after he slipped while walking without his braces.

As the double-bar caliper braces did not afford sufficient stability for ambulation without outside support, the patient was forced to rely upon crutches which lead to recurrent hemorrhages into both elbows and painful hemophilic arthropathies as a result of constant trauma to the joints

At the time of his first orthopaedic examination on January 8, 1953, the patient, then 24 years of age, was crutch ambulatory with marked hemophilic arthropathy of the knees It was interesting to observe that despite immobilization of both knee joints in complete extension by jointless braces for 15 years, a considerable range of motion had been preserved, with the right knee moving from AGE 175 degrees to AGF 90 degrees and the left knee from AGE 170 degrees to AGF 90 degrees This substantiated the principle that adequate splinting of diseased joints can prevent recurrent hemorrhages as well as flexion contractures and the tendency towards ankylosis

The left ankle was held in equinous position of 100 degrees with almost complete loss of motion, the right ankle had dorsiflexion to 90 degrees with some motion in the direction of plantar flexion

There was swelling and slight deformity of the elbows with restriction of motion at the right elbow from AGE 140 degrees to AGF 50 degrees, at the left from AGE 110 degrees to AGF 80 degrees The left elbow appeared in much worse condition with marked crepitation on passive motion and considerable restriction of pronation and supination of the forearm

After orthopaedic treatment was delayed by the patient's extended stay at a convalescent home following a nervous breakdown, he was treated ambulatory from April 23, 1953 to May 19, 1953 This was the only case in the series for which hemophilia Hessing braces were constructed with Lofstrand lock joints at the knee Use of the lock joint was indicated primarily to achieve sufficient leg stability so that crutches could be discarded thereby eliminating recurrent

trauma to the elbows. The braces immediately provided better support and the patient was soon able to walk very well without crutches except when travelling to work by bus. A last examination on July 8, 1953 indicated that the elbows were considerably improved since the patient had been able to discard his crutches In addition, quadriceps function was greatly restored, particularly on the left side, where active extension of the knee was possible almost to the point where the Lofstrand joints became self-locking, while extension of the right knee was possible to 170 degrees

Case #8182 (See Figure 35)

By the time Carl G was 17 years old, the condition of his lower extremities had been severely affected by inconsistency of diagnosis and treatment. As the boy's father was publicity director for a large organization, the family had been obliged to travel a great deal and management of Carl's orthopaedic disabilities had varied with each move.

With a family history that was negative for hemophilia, the patient was born with a hematoma that was aspirated 24 hours later. A diagnosis of hemophilia, suspected at the time, was not confirmed until the child was 10 months old This diagnosis was subsequently questioned and the patient was called a pseudo-hemophiliac.

After involvement of the left knee at 18 months of age, and of the right knee shortly thereafter, the patient was finally fitted with leg braces when he was 4 years old, following treatment of flexion contractures by means of traction At about this time, the boy suffered two cerebral vascular accidents which left him mentally retarded with spasticity of the lower extremities, microsomia of the left leg, and an equinovarus deformity of the left foot with partial ankylosis

Despite continuing use of braces and a walker, there were recurrent hemorrhages into knee joints After the patient received his last pair of braces three years ago at 14 years of age, he was restricted to bed rest and wheelchair ambulation. Rarely attending school, he relied largely upon home instruction for his education.

At the first orthopaedic examination on April 19, 1956, the patient appeared to be a friendly, cooperative and obviously retarded child with a tendency toward obesity Flexion contractures at the knees restricted motion on the right side from AGE 160 degrees to AGF 80 degrees, and on the left from AGE 145 degrees to AGF 60 degrees. The left lower-leg was considerably shorter than the right with the foot exhibiting a marked equinovarus deformity with partial ankylosis.

The patient was immediately hospitalized for complete roentgen and neurological examinations, and orthopaedic treatment of flexion contractures at the knees. Correction of the deformity of the left foot and ankle was not to be attempted.

X-ray films, taken April 19, 1956, revealed severe bilateral knee joint involvement, more extensive on the right side, with moderate soft tissue swelling around the left knee. The right foot and ankle showed moderate decalcification, with a

mild degree of hemophilic arthropathy at the ankle and some equinus position Pictures of the left foot and ankle revealed no evidence of hemophilic arthropathy but substantiated the clinical observation of deformity and ankylosis

A neurological examination confirmed the patient's mental retardation and showed the left lower extremity to be smaller than the right with the foot in equinovarus position with paralysis of dorsoflexors and hyperreflexia The left Babinski could not be tested because of the dorsiflexion of the big toe.

These examinations were followed by carefully planned orthopaedic treatment Construction of a brace that would produce correct weight-bearing alignment of the left leg and foot presented an unusually difficult problem After plaster of Paris models were made for both legs and, separately, for the left foot and ankle, a wedge cast was applied on the left side with the knee in the best possible extension of 145 degrees Thereafter, a series of wedgings obtained extension at the knee to 160 degrees, and the leg was immobilized in a well-padded plaster of Paris cylinder until the patient could be fitted with two hemophilia Hessing braces with Lofstrand spring joints at the knees for further correction of residual flexion deformities. The foot section of the left brace was specially constructed to support the body in correct weight-bearing alignment regardless of the foot's deviation into the varus position. After the patient received a pair of stock shoes, he began to stand and walk with considerable agility for the first time in more than 3 years

During this 23-day hospital admission terminating on May 12, 1956, Carl's condition was complicated by a hematuria episode which subsided, however, without transfusions Although the patient had always responded favorably to plasma and whole blood transfusions, these were indicated only in emergencies because of the poor state of the patient's veins He was finally discharged ambulatory without crutches or canes, demonstrating a tremendous improvement after a relatively short period of treatment

Case #7984

Nelson D, age 5, presented a striking example of the adverse effects of improper treatment of a hemophilic knee joint when he appeared for his first orthopaedic examination

In a family with no known history of hemophilia, Nelson was the younger brother of two girls, aged 6 and 12. Mild bleeding at birth was followed, at circumcision, by a severe hemorrhage requiring sixteen sutures Thereafter, there were many hemorrhages but none, including those into elbows and knee joints, produced permanent damage

The disability at the left knee joint that immobilized the boy and led to his first orthopaedic examination was caused by trauma sustained in a bicycle accident in June 1955 The next month, during a hospital admission for an acute appendicitis attack, the left knee became markedly swollen After transfusions, the patient made a good recovery and was discharged from the hospital in August Early in September, the left knee joint again became markedly swollen and painful Bleeding stopped after a transfusion but swelling recurred as soon as the pa-

tient attempted to take a few steps Hospitalized again from November 28 to December 20, the patient received two transfusions daily for 2 weeks The left knee joint was also aspirated under cover of transfusion A large amount of blood was removed from the joint and Hydrocortone was injected. Excruciating pain, attributed at the time to a tight strapping, subsided when the strapping was removed, but a flexion contracture at the left knee developed and was treated by traction for one week. While undergoing this form of treatment, the patient was also instructed to perform exercises During the last week of hospital treatment, there were no further hemorrhages into the left knee and no significant pain The patient was permitted to leave the hospital with instructions to walk a little each day wearing an elastic bandage on his left knee.

Thus, unprotected weight-bearing on a hemophilic joint became the final phase of ill-advised treatment which had included aspiration, injection of Hydrocortone, traction and exercises. On December 24, 4 days after he had been discharged from the hospital, he awoke with severe pain and a large hemorrhage into the left knee joint and remained bedridden until his first orthopaedic examination on December 27, 1955 At that time, the left knee joint was completely extended but moderately enlarged with some ballottement of the patella.

Roentgen examination revealed a marked increase in soft tissue density around the left knee. Except for the patella which was much larger than on the right side, all osseous structures of the knee were within physiologic limits

Treatment on an ambulatory basis was immediately initiated with application of a well-padded protective plaster of Paris cast to the left leg from toes to groin holding the knee in complete extension When this cast was removed, appearance of the joint was greatly improved and a plaster of Paris cylinder was applied to the leg in the same position.

During this period, the right foot exhibited painful swelling on the plantar and medial aspect, most marked just distal to the os calcis. A well-padded protective plaster of Paris cast was applied to the right lower-leg without attempting to change the equinovarus position of the foot. With the affected joint completely immobilized, the left knee showed marked improvement while symptoms of the recent hemorrhage at the right foot had completely subsided

Following a severe hemorrhage into the chest wall and other indications of a "bleeding cycle," the patient was hospitalized on January 10, 1956 After plasma was administered, further treatment of the left leg was considered advisable and a well-padded plaster of Paris maintenance cast was applied to the leg holding the knee in complete extension The right foot and ankle required no further treatment After a 3-day hospital admission, the patient was discharged with no evidence of continued bleeding.

On January 17, 1956, the cast on the left leg was removed and, as the knee appeared almost normal, partial immobilization on a small posterior flexible knee splint with elastic bandage was considered sufficient.

Two days later, however, the patient was again bedridden with swelling and pain at the left knee Examination revealed some tenderness and well-localized swelling over the medial aspect of the femoral condyles without significant local

heat or pain The boy's parents were warned not to be overanxious and to refrain from requesting plasma transfusions unless there was evidence of an acute hemorrhage.

A plaster of Paris cylinder was immediately applied to the left leg with the knee in complete extension and the patient was instructed to remain wheelchair ambulatory until the cast was removed. At that point, a posterior flexible knee splint was reapplied, the patient was advised to start walking as tolerated, and a Hewitt elastic knee brace was ordered

On February 3, however, the boy suffered a new hemorrhage into the left knee and complained of severe pain in the joint for which his parents had already secured two units of quick frozen antihemophilic plasma and analgesics A foam rubber-padded plaster of Paris cast was immediately applied to the leg from the toes to below the groin holding the knee at AGE 180 degrees and the ankle at AGE 90 degrees Parents were again asked to restrict the use of plasma to emergencies

For 6 weeks, attempts had been made to stop and prevent bleeding into the left knee without resorting to a large orthopaedic appliance Recurrent hemorrhages, *however, established the need for permanent protection and support of* the left knee by means of a hemophilia Hessing brace with Lofstrand spring joints Accordingly, a plaster model was made and a walking cast was applied When the appliance was delivered, the patient also received an elastic cap containing a foam rubber pad to protect the medial aspect of the right knee against contusions from the Lofstrand spring joint of the left hemophilia Hessing brace

This period of intensive treatment was concluded on March 1, 1956 By March 26, the left knee had become asymptomatic, ambulation was completely painless and the patient was permitted to start kindergarten At the end of April, he was walking very well, having been entirely free of hemorrhages at the knee joint.

When last examined on June 19, 1956, the left knee was almost normal in appearance and completely extended without lateral instability Parents of the patient were advised that he might swim during the summer and take a few steps at the beach without his appliance as long as the knee remained asymptomatic

ABSTRACTS*

Case #6809 (Age, 7)†

History: Negative family history Only child Diagnosis at age 1 Active life, attended school with occasional interruptions when laid up with hemorrhages into lower extremities

Age 4 Hemorrhages into knee joints Since then, occasional hemorrhages into or around knees. All subsided without residual deformity or disability

Age 5 Hemorrhage into right ankle which became repeatedly involved there-

* Text of the abstracts prior to our first orthopaedic examination of each patient is based on the account given by the patient or his parents

† Patient's age at time of first orthopaedic examination by the author

after. During these episodes, there was pain and swelling, and patient was unable to bear weight on right foot. He remained under treatment at a hospital where he received weekly prophylactic injections of 50 cc. plasma For three weeks prior to the first orthopaedic examination he received 100 cc. plasma per week because of pain and swelling at ankle. He was bedridden during that period No orthopaedic treatment

First Orthopaedic Examination (12/21/53): *Right ankle.* Moderate swelling but no signs of recent hemorrhage No local heat. Tenderness on palpation and pressure most marked below internal and external malleolus

Right lower-leg: Some atrophy of musculature. Pedis valgi II.

X-ray (12/29/53): *Right ankle.* Considerable increase in shadow density of soft tissues suggestive of synovitis. Apart from moderate decalcification of osseous structures, no evidence of involvement with hemophilic arthropathy.

Ambulatory Treatment (1/28/54 to 2/6/54): A small orthopaedic appliance, such as a spiral-bar lower-leg brace, was definitely indicated as unprotected weight-bearing could lead to permanent damage of right ankle. The patient, however, presented a difficult psychological problem. His mother reported that he would be greatly disturbed by having to wear an appliance, particularly one that was visible. For this reason, he received only Whitman-type foot plates for correction of valgus position of feet

Examination (7/8/54): *Right ankle.* Patient was again hospitalized, this time for five days, but received no orthopaedic treatment and was permitted to bear weight as tolerated. By the time he appeared for an orthopaedic examination, the ankle was markedly swollen with local heat and some restriction of motion. Dorsiflexion was limited to 100 degrees

X-ray (7/8/54): *Right ankle.* Increased density of soft tissue shadows of joint corresponding to recent hemarthros. Mild degree of generalized decalcification of all osseous structures at joint and distal to it was noted in comparison to left side Distal epiphysis of fibula and medial section of distal epiphysis of tibia appeared larger than same structures at left side. Enlargement of distal epiphyses of ankle was of recent origin probably due to stimulation of epiphyseal development by repeated hemorrhages into joint. Apart from these changes, there was no evidence of hemophilic arthropathy involving astragalus or other components of the joint.

Ambulatory Treatment (7/9/54 to 9/7/54): Right foot and ankle were immobilized in a well-padded cast applied from toes to below the knee. After weight-bearing was permitted as tolerated, the cast was removed and the leg was supported by 3" Ace bandage When patient complained of pain on weight-bearing, examination of right ankle revealed a very slight degree of swelling around internal malleolus, no significant restriction of motion, but a slight spastic resistance against complete supination. The Whitman-type foot plate was converted into a spiral-bar lower-leg brace by temporary addition of a light aluminum bar with riveted ankle joint and small calf band. Patient was able to stand and walk without pain

Ambulatory Treatment (5/21/55): Patient received a new spiral-bar lower-leg brace for the right foot and ankle.

Examination (4/5/56): Left elbow· Had several hemorrhages Joint was slightly swollen but without local heat and pain Motion was restricted to AGE 150 degrees, AGF 90 degrees To be fitted with fiberglass night splint for immobilization in best possible extension

Right ankle Remained completely asymptomatic as long as brace was worn There was some residual infiltration to both sides of the tendo Achillis Dorsiflexion was possible to 90 degrees.

Examination (4/28/56): Left elbow Hemophilic arthropathy.

Ambulatory Treatment (4/28/56): Fiberglass brace was made for left elbow

Case #5316 (Age, 24) (See Figure 12)

History: Uncle died of hemophilia

Age 2 First transfusion.

Age 2-13 Many transfusions for hemorrhages into joints

Age 13 Appendectomy.

Age 14 Severe hemorrhages into right knee Wore leg brace ever since

Age 18 Surgery for removal of impacted wisdom teeth

Since age 18 Hemorrhages into mesentery and abdominal walls, closing off intestines and necessitating use of Miller Abbott tubes and Wagensteins

Age 23-24 Hemorrhages into kidneys, abdominal walls, shoulders, elbows

Since age 17. Averaged 30 transfusions a year

Past 10 months 84 transfusions

Disability at first examination Crutch ambulatory wearing double-bar caliper brace on right leg with Thomas half-ring at its upper end and without knee joint

First Orthopaedic Examination (11/4/50): Right knee joint No pain or swelling Ankylosed at AGE 160 degrees with moderate degree of posterior subluxation of tibia.

Left knee joint Moderately swollen and painful Range of motion between AGE 160 degrees and AGF 130 degrees

Legs Considerable atrophy of musculature

X-ray (11/4/50): Left knee joint Considerable generalized decalcification of skeleton Articular space was fairly well preserved, particularly between lateral condyles of femur and tibia Articular space on medial aspect also well preserved but more irregular It appeared that medial condyle of femur was squared off Lateral view showed ankylosis between patella and femur

Right knee joint Greater decalcification than on left side Bones appeared markedly atrophic, particularly tibia and fibula Solid ankylosis was present between condyles of femur and tibia with good bony trabeculation running from femur to tibia and vice versa Patella firmly ankylosed to femoral condyles

Ambulatory Treatment (11/4/50 to 2/15/51): Plaster of Paris models were made for a molded leather-steel reinforced brace for right knee extending from mid thigh to mid-lower-leg, and for a hemophilia Hessing brace with Lofstrand spring joints at knee for left leg Patient was able to get up, stand and walk without a cane, made excellent progress, outlook on life changed

Last Follow-up (11/15/51): Letter reported patient's death from a cerebral hemorrhage following continuous bleeding which 37 transfusions failed to stop

Case #6264 (Age, 4)

History: Parents not certain of diagnosis.

At birth: Two large hematomas on skull despite delivery without instruments. First transfusion before leaving hospital.

5 months. Circumcision followed by hemorrhage necessitating transfusion Two incidents of hemorrhaging after injury to tongue.

April 1952: Spraining of right ankle was a frequent occurrence X-ray was negative for bone pathology.

January 1953: Right ankle suffered trauma. Unable to bear weight on right leg Cane ambulatory

Cycle. Less tendency to bleed when spending time in sun and in swimming

***First Orthopaedic Examination* (1/7/53):** Very active child; constantly jumping and pounding on feet. True or severe hemophiliac would have suffered considerable damage to ankle joints. *Evidence of many bruises on skin* with discoloration from superficial hematomas throughout body.

Right ankle joint: Enlarged with some restriction of dorsiflexion and local heat over astragalus. *Clinical* findings rather than history of patient *suggested* involvement with hemophilic arthropathy.

***X-ray* (1/10/53):** *Right ankle joint:* Some soft tissue swelling and infiltration. There was an area of decalcification and loss of structure in astragalus with possible break in cortex toward articulation with calcaneus. One film suggested an area of bone destruction due to hemorrhaging. Additional small cone views, well centered over astragalus, showed slight decalcification of area but no definite focus of bone destruction

Diagnosis: Although x-ray findings did not definitely confirm bone pathology due to hemophilia, clinical findings of local heat, restriction of motion and swelling as well as some decalcification in astragalus warranted temporary support and immobilization of right foot and ankle

***Ambulatory Treatment* (1/15/53):** Patient received molded leather boot, reinforced with a Whitman-type plate and a posterior bar extending to border of middle and distal third of right lower-leg, to be worn for 6-8 weeks.

***X-ray* (2/9/53):** *Right foot and ankle.* No definite evidence of hemophilic bone involvement

***Ambulatory Treatment* (2/11/53):** *Patient given posterior flexible knee splint* for pain and swelling at right knee following fall

***Examination* (2/16/53):** *Right knee joint* Following plasma transfusion (patient's first) on 2/11/53, knee became painless. *Swelling had disappeared almost* completely.

Right foot: Appearanc [illegible]

***Examination* (11/27/5**[illegible] 953 X-rays taken at a hospital were [illegible] ht foot and ankle Required no treatment At that time, blood tests confirmed patient's classification as true hemophiliac. On 11/17/53 patient *had developed* painful swelling of right calf, with local heat.

Examination revealed discoloration of skin from a hematoma extending down to medial malleolus and heel. Swelling and some infiltration along course of saphena magna extended toward mid-thigh

Right ankle joint: Restriction of motion with foot in equinus position of 110 degrees

Right knee joint Joint was not swollen but could not be extended beyond AGE 150 degrees.

Ambulatory Treatment (11/27/53): Bed rest, reapplication of old jointless molded leather brace for right foot and ankle to correct equinus deformity, application of posterior knee splint for right knee.

Examination (12/4/53): Right knee joint could be fully extended Swelling and induration of soft tissues on medial aspect of right calf and knee subsided Discoloration of skin limited to medial aspect of right heel and ankle Dorsiflexion of right foot improved

Ambulatory Treatment (12/4/53). Splint to be discarded Ankle brace to be worn for another week or two depending on condition of foot Patient to remain off feet until all symptoms from recurring hemorrhage into right calf had subsided

Examination (7/5/55): *Right ankle joint* Became repeatedly swollen and painful in June 1955 and patient received plasma After pain and swelling had disappeared, ankle was injured again with recurrence of pain and swelling There was marked swelling and some discoloration of skin at time of this examination and foot was held in marked equinus

Ambulatory Treatment (7/5/55 to 8/24/55): Treatment for the right ankle included successive applications of two maintenance casts, a walking cast holding foot at dorsiflexion of 90 degrees, and a 2" Ace bandage

X-ray (9/2/55): *Right ankle joint* Definite involvement with mild hemophilic arthropathy.

Articular surface of astragalus toward tibia was irregular and showed some impression of cortex The distal epiphysis of tibia had grown faster than the normal epiphysis of left tibia The soft tissues under the tendo Achilis showed increased density.

Examination (9/8/55): *Right ankle joint* Moderately swollen with some pitting edema and local heat Weight-bearing was definitely painful and to be avoided until all swelling had disappeared

Ambulatory Treatment (9/8/55): Patient received spiral-bar lower-leg brace with milled ankle joint and molded leather sandal for protection and support of right ankle to be worn day and night

Examination (4/23/56): *Left knee joint* Hemorrhage in February 1956 Received plasma at a hospital Restricted activity for 3 weeks and then permitted to return to school although knee was still enlarged. On 4/10/56 knee became stiff and increasingly swollen Ace bandage was applied. Examination revealed considerable swelling and local heat with ballottement of patella

Right knee joint Spiral-bar brace had been worn irregularly until December 1955 and then discarded Wore firm bandage on right foot and ankle.

X-ray (4/23/56): Left knee joint. Increased density of soft tissue but no bone involvement.

Ambulatory Treatment (4/23/56): Left knee immobilized and well compressed by using elastic bandage against a posterior flexible knee splint.

April 1956. Hemorrhage into left knee

LHH Treatment * *(4/28/56 to 5/8/56, 10 days):* Treatment for hemorrhages into left knee and right elbow.

Protective casts were applied to left knee and right elbow, and patient was discharged wearing casts.

Ambulatory Treatment (5/8/56 to 5/30/56): Cast on right arm was no longer necessary, left leg was splinted

Last Follow-up (5/30/56): Patient was still under treatment.

Case #7769 (Age, 23)

History: Two normal brothers; one hemophiliac brother who died in 1950, probably of a brain hemorrhage Sister's son normal.

Clinical onset. Hemophilia was discovered in infancy with severe bleeding from a minor scratch.

Preschool years· Nosebleeds

Early school years Recurrent joint hemorrhages with flexion contractures of knees and left elbow.

Age 11· Unable to walk because of flexion contractures of both knees Treated at a hospital At end of hospital admission, he was able to walk with stiff legs Limited motion gradually returned to both knees.

High school years Shoulder and elbow hemorrhages caused most difficulty.

First year of college, 1950: Severe hemorrhage into left lower back and hip resulting in inability to use left leg A left leg brace was obtained

Recent years Hemophilia was somewhat controlled. Patient was hospitalized about three times a year and treated with fresh whole blood or fresh frozen plasma. Between transfusions, there had been only relatively minor bleeding with no crippling hemorrhages since injury to left leg in 1950

1953 Internal hemorrhages in right jaw following dental work, checked by antihemophilic plasma

Cycle Hemorrhages were likely to occur in spring and fall

Diagnosis. Severe hemophiliac.

Miscellaneous clinical data. Quinsy, 1947.

Personal data Schooling. Many prolonged absences in elementary and high school Limited absences in college when hospitalized Employment throughout college years. Janitor, soda jerk, clerk Marriage: December 1954. Offspring: None. Activities. Played golf regularly with little difficulty.

Hospital treatment. Hospitalized about 25 times, mostly in 5 years prior to first orthopaedic examination Average admission was 3 to 6 days. Received one or two transfusions per hospital admission Whole blood, to which patient appeared to respond better, was given more often than fresh frozen plasma.

* Lenox Hill Hospital admission

Orthopaedic treatment: Patient had worn a double-bar left leg brace for many years, for stabilization of knee joint

First Orthopaedic Examination (8/30/55): Patient was cane ambulatory and in good general condition.

Left knee joint: Showed involvement with hemophilic arthropathy of long standing Although patient could take a few steps without brace, he usually stood and walked with joint of brace locked in complete extension Had rather well developed quadriceps and was able to move joint actively from AGE 170 degrees to AGF 110 degrees No swelling or pain.

Right knee joint: Showed involvement with hemophilic arthropathy of long standing Recurrent hemorrhages had affected joint which had not been braced Moderate swelling and pain due to recent mild hemorrhage throughout lateral aspect.

X-ray (8/30/55): *Left knee joint* Moderate degree of hemophilic arthropathy involving all structures of joint Appearance of knee indicated involvement early in life with characteristic deformity of patella more marked than on right side. There was also a rather large osteochondroma originating from posterior aspect of external condyle of femur

Right knee joint. Moderate degree of hemophilic arthropathy involving all structures of joint Some soft tissue swelling, which was not present on left side Articular space was better preserved than on left side

Ambulatory Treatment (8/30/55): A plaster of Paris model was made of left leg to be used in manufacture of hemophilia Hessing brace

Examination (8/31/55): *Left shoulder.* Some restriction of motion, with elevation and abduction possible to 110 degrees

Right elbow. Slight restriction of complete extension from a small hemorrhage in July 1955. Appearance and function of right arm normal in every other respect

Left elbow. Involvement with hemophilic arthropathy, with some deformity and restriction of motion from AGE 140 degrees to AGF 50 degrees There was almost complete loss of supination of forearm, while pronation was hardly restricted.

Right knee joint Swelling had subsided considerably.

Ambulatory Treatment (9/7/55 to 9/12/55): Patient received Hewitt-type elastic brace for right knee, which offered all necessary support, and a hemophilia Hessing brace for the left leg with Lofstrand spring joints at the knee, ankle joint and sandal After delivery of these appliances, which were well tolerated, patient was considered completely rehabilitated from an orthopaedic point of view.

Last Follow-up (3/25/56)· Patient died of massive abdominal hemorrhage

Case #7146 (Age, 8¾)

History No definite family history, mother's father may have been a hemophiliac. One sister, 11 years old.

Circumcision· Abnormal bleeding

Age 18 months: Patient cut upper lip, hospitalized for two weeks, bleeding stopped by topical applications

Age 22 months: Hemophilia diagnosis.

Age 5 years. Contusion to left ankle with hemorrhage into joint, 4 transfusions.

Age 6 years. Kidney hemorrhage; one transfusion Contusion to right elbow, followed by hemorrhaging. Since then, elbow had been stiff occasionally but mostly freely movable

Recent years. Three injections of 50 cc Hyland plasma as prophylaxis when patient had stomach flu Tendency toward bruising was corrected for only one or two days after injections

Complaints: Left ankle painful and swollen after increased activities. Codeine had been administered for pain but not during past year Tendency toward pain and swelling at right ankle attributed to weight-bearing on right leg.

Schooling Able to attend, when left ankle became painful or swollen, he was carried.

***First Orthopaedic Examination* (8/17/54):** Good general condition, bedridden for past two months.

Left ankle and foot: Moderate deformity with thickening of capsule and entire region of tendo Achillis, which was less prominent than on right side. There was no significant restriction of motion at ankle and foot but crepitation originated from joint proper

Left leg: Very little muscular atrophy.

Right ankle and foot: No swelling or deformity of ankle; foot in moderate valgus position.

Right arm. Some atrophy of musculature.

Right elbow: AGE 150 degrees; AGF 50 degrees.

Left elbow. AGE 180 degrees, AGF 60 degrees

***X-ray* (8/17/54):** *Left ankle and foot:* Generalized decalcification of osseous structures more marked than on right side. Left ankle showed definite involvement with hemophilic arthropathy with flattening of superior articular surface of astragalus, small juxta-articular cysts in distal epiphysis of tibia and some thinning of articular cartilage. Tibia and fibula appeared considerably thinner than on right side.

Right foot and ankle: Generalized decalcification, less marked than on left side No bone pathology but definite thickening of ankle joint capsule, suggestive of previous hemorrhages into joint.

Diagnosis: *Left ankle.* Moderate hemophilic arthropathy though function of left foot and ankle was quite good

Right ankle Mild hemophilic arthropathy.

Right elbow Mild hemophilic arthropathy.

***Ambulatory Treatment* (8/19/54 to 8/31/54):** Patient was fitted for and received a small Hessing-type brace for the left lower-leg with milled ankle joint, and a fiberglass Whitman-type foot plate for the right foot, both made to plaster of Paris models. Examination on 8/31/54 revealed *pain at right ankle* that pre-

vented weight-bearing on right foot. There was considerable swelling between internal and external malleolus and tendo Achillis Patient was fitted with small elastic stocking to control swelling. This was not tolerated as it increased pain

Ambulatory Treatment (9/3/54 to 9/6/54). A protective cast was applied to right foot and lower-leg to speed healing process After cast was removed, the Whitman-type foot plate was reapplied and patient was permitted to walk

Examination (12/20/54): Patient had walked very little.

Right ankle: No swelling of right ankle, no restriction of motion, moderate valgus

Left foot and ankle Appeared improved

Ambulatory Treatment (12/20/54 to 12/28/54): For recurrent disability at right ankle, patient received a small Hessing-type brace for the lower-leg, made to a plaster of Paris model.

Examination (6/20/55). Patient walked very well

Left ankle. Some soft tissue swelling following recent hemorrhage

X-ray (6/20/55): *Left ankle* Some increased soft tissue swelling

Feet Osseous structures appeared better calcified

Ambulatory Treatment (6/29/55 to 7/14/55): Patient received new Hessing-type braces, made to plaster of Paris models, for both lower-legs and feet Permitted to swim.

X-ray (10/24/55): (Films were mailed from an out-of-town hospital) *Right elbow.* Moderate degree of hemophilic arthropathy from injury at six years of age. Bone involvement chiefly limited to ulnar region of articular and juxta-articular section of distal end of humerus. Epiphysis of head of radius measured 19 mm in comparison to 17mm on left side showing usual overgrowth in earlier affected articulation. The olecranon showed some deformity Articular space was otherwise well preserved Calcium density of osseous structures was satisfactory and there was no increased soft tissue shadow

Left elbow: Increased soft tissue density, moderate decalcification of all osseous structures in comparison with right side, some structural irregularities in trochlea of humerus, and calcification close to tip of olecranon Articular space was still well preserved

Diagnosis· Right elbow Old hemophilic arthropathy of moderate degree

Left elbow: Recent hemophilic involvement limited to soft tissue structures except for involvement of trochlea.

Examination (12/29/55): *Right elbow·* Completely asymptomatic

Left elbow· Recurrent superficial hemorrhages AGE 140 degrees, AGF 90 degrees.

Ankles· Asymptomatic with definite clinical signs of arthropathy

Case #6271 (Age, 16) (See Figures 2 and 32)

History: Hemophilia in family; brother age 18, two maternal uncles, cousin (died).

4 months· First hemorrhage

Joints Moderate hemorrhages.

11/11/52: Accident to right knee; unable to bear weight on leg

1/7/53: Last transfusion for hematuria

School: Did not attend although he was ambulatory until 11/11/52 Home tutor.

First Orthopaedic Examination (1/10/53): Reduced general condition; pale, bedridden.

Right knee joint: Moderate swelling and deformity, no fluid within joint Joint and proximal third of tibia extremely tender to pressure and to slight passive motion, AGE 135 degrees.

Right ankle. Swollen and rigid.

Right leg· Marked atrophy of musculature.

Left knee joint· AGE 170 degrees; no pain.

Left leg· Extensive muscular atrophy.

X-ray (1/17/53): Right knee joint· Involvement with moderate hemophilic arthropathy with thinning of articular space, irregularity of articular surfaces, most marked on medial aspect of knee Considerable juxta-articular pathology noted in internal condyle of tibia.

Films indicated that injury on 11/11/52 caused fracture of upper portion of tibia about 2½ inches below knee without significant displacement or angulation Fracture lines were still visualized but there was solid union. An almost completely healed fracture of the fibula was noted at the same level

Right foot and ankle· Marked spotty decalcification of all osseous structures and considerable thinning of cartilage of ankle joint and tarsal articulations

Left knee joint: Well marked deformity of patella with squaring off and loss of distal pole Some thinning of cartilage on medial aspect.

***LHH Treatment* (1/17/53 to 2/27/53, 41 days):** During this hospitalization, patient was treated for hemophilic arthropathy of right knee with flexion contracture of 135 degrees, residual fracture of proximal end of tibia and fibula and for reflexdystrophy of right foot and ankle. Preliminary treatment of right knee consisted of complete bed rest, immobilization of knee on posterior flexible knee splint, and refrigeration A wedge cast was then applied. This cast was removed with knee held in extension of 165 degrees and a plaster of Paris model was made of the leg to be used in the manufacture of a hemophilia Hessing brace with Lofstrand spring joints at the knee and simple milled ankle joints The procedure was performed under Avertin anaesthesia supplemented by gas After model was taken, a maintenance cast was applied to the right leg, holding knee at AGE 165 degrees and ankle at AGE 110 degrees. During this time, no attempt was made to correct the equinus position of the right foot. The right ankle was no longer swollen.

Patient was discharged crutch ambulatory wearing a hemophilia Hessing brace on his right leg, holding knee in extension of 165 degrees

Examination* (3/26/53): *Right knee joint: Completely extended, no swelling, continuing weight-bearing pain throughout right leg due to reflexdystrophy.

Patient remained crutch ambulatory as he walked very poorly without crutches and was unable to stand erect Left shoe was fitted with lift to compensate for

apparent increase in length of right leg with brace and knee held in extension

Examination (4/2/53): Right foot Large area of discolored skin and threatening decubitus over posterior aspect of heel and tendo Achillis

Examination (5/14/53): Left shoulder Soft tissues of entire upper arm were under considerable tension with local heat and restriction of motion due to recent acute hemorrhage into shoulder and upper arm

Right foot. Still extremely tender, entire right lower-leg, ankle and foot showed considerable atrophy Decubitus on posterior aspect of heel was greatly improved.

Examination (5/27/53): Left shoulder and upper arm. Marked swelling and infiltration due to repeated hemorrhages Biceps was mostly affected Arm was held close to trunk and there was very little active or passive motion without producing pain

Right foot Decubitus on heel was almost healed

LIIII Treatment (5/28/53 to 6/11/53, 14 days): For acute hemorrhages into left shoulder and upper arm due to repeated trauma from crutches

Examination (5/28/53). Left shoulder Markedly swollen, very little range of motion Arm was held close to trunk There was some local heat and extensive discoloration of skin from hematoma

Right leg: Tendency toward equinovarus position of foot, skin was in poor condition due to trophic changes

X-ray (5/28/53): Left shoulder. Films showed a very thin shaft of humerus but no involvement of shoulder joint proper and juxta-articular structures with hemophilic arthropathy

Right foot and ankle Moderate but definite degree of recalcification with better defined structural details.

During this hospital admission, application of ice to left shoulder was followed by placing of shoulder on an abduction aeroplane splint until acute symptoms subsided Patient was to avoid use of crutches

For right lower-leg and foot, whirlpool baths and underwater exercises were prescribed to decrease skin sensitivity and improve nutrition of soft tissues General condition was greatly improved

Examination (6/18/53): Right foot and ankle. Decubitus on heel was completely healed. Skin showed definite improvement Trophic changes were less marked and foot was less sensitive to light touch There was slight active motion in direction of plantar flexion and supination

Left shoulder. No pain but considerable soft tissue swelling and infiltration throughout anteromedial aspect of proximal third of upper arm, due to residual hematoma Infiltration restricted motion above shoulder level while internal rotation was almost zero

Examination (6/30/53): Ambulatory without crutches

Examination (7/28/53): Right ankle and foot Less sensitivity throughout skin, some increase in motion of ankle and of toes.

Left shoulder. Painless, freely movable, not swollen with exception of small area of soft tissue infiltration on flexor side of upper arm beneath axilla

X-ray (7/28/53): Right foot and ankle: Slight recalcification.

Examination (8/25/53): Considerable progress was noted for the first time since treatment was begun Patient walked with one cane.

Left shoulder. Asymptomatic

Right knee joint: No swelling.

Examination (9/22/53): Right knee joint. 20 degrees of flexion.

Right foot and ankle· Less trophic changes, no tenderness on palpation and pressure.

Patient was able to walk without cane

Examination (1/14/53): Hospitalized for 6 weeks for hemorrhages resulting from tooth extraction, many plasma transfusions, good recovery.

Right lower-leg and foot Much better nourished, improved range of motion at ankle.

X-ray (1/14/54): Right foot and ankle: Generalized decalcification but considerably greater calcium density, showing that reflexdystrophy which followed fracture of tibia was about to heal.

Mount Sinai Hospital Treatment (2/12/54 to 2/19/54): Admission for kidney hemorrhages. Treatment consisted of 3 plasma transfusions

Examination (3/16/54): Walked well.

Right leg Original trophic changes of skin at right ankle and foot had disappeared. Equinus position of foot remained with some return of active plantar flexion.

Examination (5/13/54): Right ankle and foot Foot showed further improvement, motion at ankle increased by a few degrees

Recommended. Addition of strong crossed elastics connecting distal margin of sandal with 2 studs on lower-leg section of hemophilia Hessing brace

7/13/54 to 11/11/54: Internal hemorrhages

Examination (5/5/55): Right knee joint Attempts to bend joint were without success

Right lower-leg· Musculature strong and plantar flexion excellent but dorsiflexion of foot and ankle was still weak and restricted.

Recommended· Exercises to improve dorsiflexion, no attempt to be made to bend right knee Patient to take a few steps at home without brace. Aside from *this,* appliance to be worn *continuously*

6/2/55 to 12/1/55: Repeated Mount Sinai Hospital admissions for hemorrhages which required transfusions

Examination: Right knee joint Asymptomatic.

Ambulatory Treatment (1/23/56 to 2/6/56): Construction of a new hemophilia *Hessing brace for the right leg*

Last Follow-up (July 1956)

After complete orthopaedic rehabilitation with an appliance for the right leg, the patient suffered hemorrhages into the left knee starting in April 1956, and into the left hip region in May 1956 Because of hemophilic arthropathy of the *left knee, he was unable to lift the left leg from a supine* position and was forced to give up his job as a sorter of instrument parts He required a hemophilia Hessing brace for the left leg.

Case #5624 (Age, 2) (See Figure 1)

History: Only child, no known hemophilia in family.

Circumcision: No abnormal bleeding

Since 9 months of age Bruised easily.

Age 15 months: First hemorrhage into left knee joint which became painful and enlarged, with a flexion contracture of 90 degrees.

Age 17 months Right knee exhibited swelling for 5 days.

May 1951 to July 1951: Persistent swelling of left knee, patient was bedridden

LIHI Treatment (7/16/51 to 8/10/51, 25 days): For early stage of hemophilic arthropathy and synovitis at left knee

First Orthopaedic Examination (7/17/51): Well-developed child, numerous hematoma and ecchymoses on trunk and all extremities

Left knee joint: Considerable soft tissue swelling and enlargement No definite evidence of free fluid Knee held in flexion of approximately 100 degrees Range of motion restricted to AGE 100 degrees, AGF 90 degrees

X-ray (7/17/51): *Left knee joint* Mild degree of generalized decalcification of osseous structures Pathologic changes suggestive of early involvement with hemophilic arthropathy were limited chiefly to soft tissues Marked density of capsule was noted There was possibility of fluid within joint. Articular space showed some distension in comparison with right side Structural irregularities of distal femoral epiphysis were most marked on medial aspect

During hospitalization, a hip spica was applied under general Avertin anaesthesia to left leg from toes to distal ribs, with knee held in extension of 120 degrees. Cast was well padded in preparation for wedging at left knee X-ray, taken on 8/8/51 after wedging of cast, showed AGE 165 degrees with slight degree of posterior subluxation of tibia in relation to axis of femur. Degree of generalized decalcification was very moderate Soft tissue swelling and density of soft tissue shadow were definitely decreased since 7/17/51 After the wedge cast was removed, a maintenance cast was applied, holding knee in corrected position of AGE 165 degrees until a double-bar leg brace without knee joint and with caliper to shoe was delivered Brace was fitted with well-padded knee cap with solid leather straps to be tightened to obtain additional 15 degrees of extension over a long period of time To prevent irritation of knee joint from pressure of knee cap, a second well padded knee cap was made with elastic straps to be worn at night and whenever knee joint became irritated Patient started to walk on last day of this hospital admission

Last Follow-up (9/7/56): A letter reported that the child had been hospitalized twice in the past five years and had received three transfusions for a cut in the throat. The patient appeared to bruise at certain times of the month, walked with a slight limp, played with other children and attended second grade He had lost no time at school because of hemophilia

Case #6504 (Age, 10)

History: Mother's brother died of hemophilia.

Circumcision. Profuse bleeding.

Infancy. Cut lip; diagnosis established.

Age 16 months. Began to walk. Since then, patient has had trouble with both knee joints. Unable to walk at times for a week or two because of pain and swelling at knees.

Age 2 years. Hernia operation with many transfusions.

Age 3 years. Tooth extraction with transfusion.

Since age 3. No transfusions, no special medical care.

November 1952. Large hematoma on left knee after fall.

Since January 1953. Unable to walk because of pain at left knee.

February 1953. Left knee aspirated to relieve painful tension; improved.

Other joints involved. Left elbow. Swollen for a year with deformity and restriction of motion. Attributed to injury while wrestling. Left ankle.

Schooling. Home teacher, good student.

First Orthopaedic Examination (6/4/53): Well developed boy, unable to walk.

Left elbow. Enlarged, restriction of motion from AGE 150 degrees to AGF 70 degrees. Forearm showed no restriction of motion.

Left knee joint. Moderately swollen; contained some fluid, skin on dorsum showed striae indicating that swelling was considerably greater. AGE 150 degrees, AGF 60 degrees. Marked valgus position and moderate posterior subluxation of tibia. Acute hemophilic arthropathy.

Left ankle. Enlarged, some restriction of dorsiflexion.

X-ray (6/9/53): Left elbow. Moderate deformity. Distal end of humerus and proximal end of ulna and radius were larger than same structures at right side. Epiphysis of radial head in particular was more than twice the size of normal right epiphysis. All other ossification centers seemed to be slightly larger than on right side. There was considerable soft tissue density around the elbow. A small separate ossification was noted at tip of olecranon. Not possible to decide if this represented a separate ossification center or an incomplete fracture. The former was more probable. Articular surfaces of ulna and medial humeral condyle showed some irregularities while articulation between radius and humerus was well preserved.

Left knee joint. Marked distension of the capsule probably due to fluid within joint. Distal end of femur showed considerable enlargement in contrast to the rather thin femoral shaft. Femoral condyles and articular surface of patella as well as articular surface of tibia showed considerable irregularities due to involvement with hemophilic arthropathy. Small cystic areas existed throughout proximal epiphysis of tibia. Tunnel view showed that articular space was fairly well preserved and that articular surfaces of femur and tibia were fairly congruent.

Independent of involvement with hemophilic arthropathy, an osteochondroma was noted originating from posteromedial aspect of tibia as usual about one inch below proximal epiphysis.

Right knee joint Very slight structural irregularities, particularly at medial femoral condyle indicating involvement with hemophilia

Left ankle Generalized decalcification of mild degree, considerable thinning of articular space of joint proper, moderate overgrowth of distal epiphysis of tibia on medial side, some overgrowth of external malleolus Axis of joint showed definite displacement in direction of valgus

LIHI Treatment (6/29/53 to 7/11/53, 12 days): For deformity and flexion contracture of left knee

During hospitalization, a Quengel cast was applied from toes to below the groin which obtained complete extension of the left knee, without increase in valgus position or posterior subluxation of tibia in relation to femur After patient was fitted with a hemophilia Hessing brace with Lofstrand spring joints at the knee, simple ankle joints and sandal holding knee in complete extension, he began to walk

Ambulatory Treatment (8/25/53)· Patient received posterior flexible splint for protection of right knee

X-ray (9/26/53): *Left knee joint* AGE 175 degrees There was still considerable enlargement of soft tissues of joint with rather marked density

Examination (10/24/53): *Right knee joint* Hemarthrosis without significant local heat or pain

Left knee joint Asymptomatic, marked lateral instability with knee completely extended

Ambulatory Treatment (10/24/53)· *Right knee joint* Splinting and transfusion

Examination (12/5/53) Laceration at thenar of left thumb Three sutures

Right knee joint Patient to discard splint as long as swelling and pain did not recur

Left knee joint Lateral instability still quite marked Hemophilia Hessing brace to be worn constantly

Examination (4/15/54): *Knee joints* Moderate enlargement and apparent ballottement of patella Lateral instability of fully extended left knee almost insignificant Patient required Hewitt-type elastic knee brace for right side

X-ray (4/15/54): *Left knee joint* Less soft tissue swelling and better calcification of osseous structures without further evidence of destruction

Right knee joint Some soft tissue swelling in region of upper quadriceps pouch

Ambulatory Treatment (4/22/54). Patient received a Hewitt-type elastic brace fitted with solid steel bar in back for right knee Walked extremely well

Examination (9/18/54). *Left knee joint* Acute hemorrhage into joint which did not respond to ice Marked enlargement with ballottement of patella, local heat and pain Marked lateral instability noted

LIHI Treatment (9/18/54 to 9/25/54, 7 days). For acute hemorrhage into left knee joint Patient made complete recovery following one transfusion with quick frozen plasma, bed rest and application of ice The hemophilia Hessing

brace was worn constantly during treatment. During this admission, patient developed a fever without other symptoms.

LHH Treatment (10/30/54 to 11/18/54, 19 days): For treatment of acute hemorrhage into right knee joint, a hemorrhage into the left elbow; and an infection of unknown origin

During this admission, patient's left elbow was treated with refrigeration and an Ace bandage, and the hemophilia Hessing brace for the left leg was reconstructed.

Examination (2/12/55): Patient suffering from recurrent attacks of tonsillitis and laryngitis.

Ambulatory Treatment (3/26/55): Patient received a new Hewitt elastic brace for the right knee.

Hospital Admission (July 1955): Patient required several whole blood and plasma transfusions during a three weeks' admission for rectal hemorrhages

Examination (10/1/55): *Left knee joint. Remained enlarged with considerable soft tissue infiltration and induration.* Definite lateral instability with knee completely extended.

Right elbow: Slight restriction of extension.

Ambulatory Treatment (10/1/55 to 12/15/55): Patient was fitted for and received a new hemophilia Hessing brace for left leg.

Last examination (3/13/56): Patient's general condition was good and he had suffered no additional hemorrhages He walked comfortably and wished to attend school.

Case #5672 (Age, 42) (See Figure 19)

History: Negative hemophilic ancestory.

Clinical onset· College years. Hemophilia did not interfere with athletic activities Years ago, patient played baseball regularly, pitching with left hand Hemophilia, never of great severity, became less and less active with advancing years and disability on locomotion, to which patient had become well adjusted, was caused chiefly by hemophilic arthropathy of both knee joints. Hemophilic elbow joints caused some disability but not enough to require active therapy. There had always been pain in knees, left elbow and occasionally in right elbow. At times, patient had suffered from attacks in which joints were hot as well as painful

Occupation. Industrialist, patient traveled a good deal using a folding wheelchair

First Orthopaedic Examination (9/27/51): *Knee joints·* Patient was confined to wheelchair as he was unable to stand or walk because of flexion deformity of knees Joints were fusiform No evidence of fluid. Considerable crepitation present on active and passive motion which was possible between AGE 150 degrees and AGF 100 degrees

Right ankle Some restriction of dorsiflexion and evidence of recent subcutaneous hemorrhage in region of internal malleolus

Right thigh: Femur showed deformity and some shortening from malunited fracture of shaft 20 years earlier

Musculature of legs. Fairly well preserved.

Elbows Marked enlargement and deformity of left elbow with restriction of extension to 125 degrees and of flexion to 85 degrees. Similar findings, to a much lesser degree, were present at right elbow with ACE 140 degrees and AGF 60 degrees

X-ray (9/29/51): *Right elbow.* Advanced stage of hemophilic arthropathy General calcium density of skeleton was normal Articular space was so thin as to indicate almost complete destruction of cartilage Head of radius and coronoid process of ulna, and to a lesser degree the olecranon, showed considerable broadening and lipping. Juxta-articular cysts were noted particularly in head of radius and lateral condyle of humerus

Left elbow· Unusual degree of productive deformity of joint. Thinning of cartilage less marked than on right side. Productive changes, particularly of coronoid process, head of radius and olecranon, were grotesque. Irregular increased calcium density throughout ulnar condyle of humerus with considerable broadening of condyle Condyles of humerus in its sagittal diameter had been worn down to thickness of shaft of humerus. They were held deep between the articular surfaces of ulna, which extended up to shaft of humerus like the shoes of a brake Marked periosteal ossification noted on dorsal aspect of proximal end of ulna

Knee joints Generalized decalcification. Marked thinning of articular cartilage with irregularities of articular surfaces of femoral and tibial condyles Articular space was somewhat better preserved on right side Right knee showed result of an old fracture of distal shaft of femur which had united with some over-riding and angulation but with solid bony union Patella was plump with considerable broadening along distal margin Few cystic changes were noted juxta-articular in medial condyle of right femur Left knee showed considerable decrease of distance between patella and femoral condyles There was also a marked deformity of medial condyle of tibia which suggested a previous breakdown, possibly due to development of juxta-articular cysts Articular surface of external tibial condyle showed 30 degrees of downward inclination The corresponding medial condyle of the femur had adjusted itself to angulation of the tibial plateau.

Right ankle Moderate involvement with thinning of articular space and irregular contour of articular surfaces

Ambulatory Treatment **(10/1/51 to 10/7/51):** For pain at knee joints from long-standing hemophilic arthropathies

As patient wanted treatment limited to the relief of pain at night, posterior celluloid splints were made to plaster of Paris models of both legs These were successful in preventing motion and pain at night

Case #6819 (Age, 10½)

History: Only child. Negative family history

9 months Hemophilia diagnosis established with hemorrhage into right knee joint Since then, knee had always shown some restriction of extension

Occasional hemorrhages into other joints no residual deformity or disability

Occasional internal and muscular hemorrhages

Age 3 Orthopaedic treatment for flexion deformity of right knee joint Not corrected

March 1953. Trauma to right knee. Disabled for many months. Unable to bear weight on right leg

October 1953· Started to walk with right knee in flexion and right-sided limp

***First Orthopaedic Examination* (1/19/54):** Well developed boy.

Right knee joint No evidence of soft tissue swelling or ballottement of patella. Flexion contracture with AGE 150 degrees, AGF 60 degrees. Definite valgus tendency.

Right leg: Musculature showed considerable atrophy.

Right foot. Narrower than left foot although of equal length

Right ankle· Slight involvement, with restriction of supination

X-ray films of right knee joint, taken on 12/1/53 at another hospital, were also inspected at this time. Skeleton of the knee showed moderately advanced changes from hemophilic arthropathy with irregular articular surfaces of femoral and tibial condyles, generalized decalcification; small juxta-articular cysts, diminished articular space, increased soft tissue density Ossification center of apophysis at tuberosity of tibia was very much larger than on left side Patella showed some changes but no squaring off at distal pole

***Ambulatory Treatment* (1/28/54 to 5/18/54):** Patient received hemophilia Hessing brace with Lofstrand spring joints at knee, milled ankle joints and elastic attachments Knee was held in extension of 155 degrees Further extension to be accomplished by means of an additional elastic attachment to be worn for two 1-hour periods each day

***Examination* (7/6/54):** *Left palm.* Hemorrhage after contusion necessitating transfusions Part of hematoma at hypothenar was aspirated with Wydase® injected Procedure was not successful. There was considerable swelling and induration most marked in region of hypothenar and on volar aspect of 5th finger. Motion at fingers was returning.

***Examination* (1/18/55):** *Right knee joint·* Occasional pain, no significant findings, AGE 175 degrees

Left ankle· Some swelling due to an old hemorrhage, and slight restriction of motion in all directions Left foot was in marked plano valgus position

***Ambulatory Treatment* (1/18/55 to 6/29/55):** Patient received a Lange-type fiberglass foot plate for left foot Right leg was so improved that springs were removed from brace joints Patient was given a Hewitt-type elastic knee brace for use during summer, as tolerated, but was instructed not to discard hemophilia Hessing brace permanently

Case #5732 (Age, 33) (*See Figure 11*)

History· Married, no children, salesman. Maternal uncle and second cousin (mother's side) died of hemophilia. Not circumcised

Age 2 Accident to forehead, hemophilia diagnosis established, wound took 5 years to heal.

Joints. Knees and elbows with deformity and functional disability.

Internal. Kidney, intestinal

Age 22 Tooth extraction necessitated 5 transfusions at hospital to stop bleeding.

Cycle. Occasional bleeding every 4 to 6 months With increase in age, hemorrhages have decreased in intensity and have occurred at greater intervals During past 6-7 years, there were no significant internal hemorrhages Joints, however, became more painful and disability on walking increased

Orthopaedic treatment Elastic knee caps for both knees

Schooling Started at age 7.

First Orthopaedic Examination (11/6/51): Patient was crutch ambulatory, holding his knees stiff and flexed Right heel did not touch ground

Legs Moderate muscular atrophy

Right knee joint No swelling or pain Motion between AGE 165 degrees and AGF 110 degrees Locking of the joint was probably due to a loose body or torn internal semilunar cartilage

Left knee joint No swelling or pain Motion AGE 150 degrees, AGF 85 degrees

Right hip Slight flexion contracture

Right ankle and foot Dorsiflexion at ankle was limited and foot was held in slight equinus position

Elbows. Moderate deformity with extension limited to 135 degrees on right side and 145 degrees on left side Flexion was almost normal Pronation and supination of forearm was restricted to 50 per cent in both directions on each side

X ray (11/7/51): *Knee joints* Almost symmetrical involvement with advanced changes characteristic of hemophilic arthropathy Extensive destruction of articular cartilage throughout femoral condyles and tibial plateau Articular space between patella and femoral condyles was markedly diminished, slightly better preserved at right knee than at the left Both patellas showed large cystic areas of bone destruction Films of knee joints revealed many juxta-articular areas of cyst-like bone destruction Pathologic changes were slightly more marked at left knee than at right

LHH Treatment (3/17/52 to 3/28/52, 11 days): For correction of flexion deformities of right and left knees due to involvement with severe hemophilic arthropathy of long standing

After plaster of Paris models were made of both legs for manufacture of hemophilia Hessing braces with Lofstrand knee joints, a wedge cast was applied to the left leg, holding the knee at AGE 145 degrees Cast was wedged three times until AGE 165 degrees was obtained After braces were fitted, an attachment for elastics was added to the appliance for the left leg in an effort to achieve greater extension at the knee, and patient was discharged ambulatory with no further pain on either side

Examination (7/21/52): Patient able to work without interruption and drive long distances without difficulty

Examination (3/21/53): Gain of fifteen pounds was attributed to freedom

from pain. Braces were worn only on long distance trips or when patient's work entailed considerable standing or walking

Right knee joint AGE 175 degrees with slight posterior subluxation of tibia, AGF 105 degrees.

Left knee joint. AGE 160 degrees, AGF 90 degrees.

Musculature of both legs was stronger.

Last Follow-up (3/23/55): Legs stronger, hemophilia Hessing braces had not been worn except on business trips or when patient was required to stand for long periods of time

Comment: Articular and juxta-articular destruction was so extensive that it was difficult to understand how knee joints could function so well without pain

Case #5566 (Age, 14)

History: Known hemophiliac since birth, considerable family history. Throughout life, had been treated at clinic where he received a number of transfusions.

Internal hemorrhages. One intracranial hemorrhage in 1947, one episode of "kidney crystals"

1948. After injury to right knee, there were frequent attacks of hemarthros, and a flexion contracture developed.

Prior to first orthopaedic examination· Some bleeding into left knee joint with beginning flexion contracture Disease appeared to be cyclic with hemorrhages occurring at various intervals with or without trauma, followed by periods of several months without bleeding. Most bleeding occurred into joints, subcutaneously, muscularly, and into mucous membranes.

First Orthopaedic Examination (6/5/51): Well developed; slightly obese; in good general condition, wheelchair ambulatory.

Right knee joint. Enlarged, moderate hemophilic arthropathy, AGE 155 degrees, AGF 115 degrees

Left knee joint. Considerable enlargement due to recent hemorrhage with flexion contracture of 150 degrees, AGF 125 degrees.

X-ray (6/5/51): *Right knee joint·* Considerable decalcification of skeleton, most marked throughout epiphyses and patella. Articular cartilage showed marked thinning and articular surfaces of femoral and tibial condyles were quite irregular There were some cystic changes throughout anterior portion of tibial epiphysis

Left knee joint No bone involvement Articular surfaces were smooth and almost congruent. Joint appeared enlarged with increased shadow density of soft tissue structures corresponding to recent hemarthros Shaft of tibia showed structural irregularities.

LIIH Treatment (6/5/51 to 6/29/51, 24 days): Protective cast was applied to left leg and swelling subsided rapidly. When cast was removed, extension at left knee reached AGE 170 degrees and a Hewitt elastic knee brace was applied. This brace controlled swelling and gave sufficient support to prevent recurrence of flexion contracture. Patient received a hemophilia Hessing brace with Lof-

strand spring joints for the right leg Extension at knee joint increased steadily and patient was discharged ambulatory without crutches or canes

Examination and Ambulatory Treatment (6/30/51): No complaints Swelling of left knee had subsided and right knee remained quiescent Patient received splint for right leg to be used instead of hemophilia Hessing brace at night.

Case #6678 (Age, 9)

History: Known hemophiliac, brother, age 15, is not

Circumcision: Bleeding.

Age 7 months Hemophilia diagnosis established when hospitalized for treatment of osteomyelitis at external malleolus of left ankle, hemorrhage occurred when vein was exposed for transfusion

Age 7-9 years Involvement of left knee joint, flexion contracture, unsuccessful attempts to correct by means of physical therapy and traction, largely crutch ambulatory

Age 9 years Hemorrhage into right knee joint after injury, flexion contracture. Other joints involved Right elbow, right shoulder, left elbow. Internal hemorrhages Chest

First Orthopaedic Examination (10/6/53): Patient had been bedridden for several months

Left elbow Some residual enlargement and stiffness

Left knee joint Moderately advanced deformity, AGE 90 degrees

Right knee joint Moderate swelling, no local heat or tenderness, AGE 100 degrees

X-ray (10/6/53): *Left elbow* Mild hemophilic arthropathy

Right elbow Soft tissue swelling

Left knee joint Moderately advanced hemophilic arthropathy

Right knee joint. Rather well preserved articular space and slight bony changes, marked soft tissue swelling and density.

LHH Treatment (10/6/53 to 10/15/53, 9 days): For correction of flexion contractures at both knee joints

During hospital admission, a Quengel cast double hip spica was applied from lower ribs to toes Subluxation hinges were incorporated at knees to correct posterior subluxation of tibia When cast was finally removed, extension at right knee had been increased by 70 degrees to AGE 170 degrees and at the left knee by 40 degrees to AGE 130 degrees A plaster of Paris model was made of right leg for a hemophilia Hessing brace with Lofstrand spring joints, and a maintenance cast was then applied to the leg On the left side, a new wedge cast was applied holding the knee in extension of 130 degrees Patient was discharged crutch ambulatory wearing two casts

Ambulatory Treatment (10/15/53 to 12 1 53). The hemophilia Hessing brace for the right leg was fitted and delivered and a plaster model was taken for a hemophilia Hessing brace for the left leg After a third wedge cast was applied to the left leg and wedged until the knee reached AGE 150 degrees, a mainte-

nance cast was applied. *This was followed by splinting until delivery of the hemophilia Hessing brace with Lofstrand spring joints temporarily locked at 150 degrees* Patient also received an attachment for elastics to be used for further correction of residual flexion contracture of left knee. He continued to be crutch ambulatory.

Examination (12/19/53): Patient discarded crutches and walked very well with his braces

Examination (1/19/54): Left knee joint. AGE 165 degrees.

Examination (4/12/54): *Left elbow.* Hemorrhage in January 1954, considerable muscle spasm throughout arm but no pain and no significant swelling or local heat. AGE 100 degrees.

Right knee joint. Asymptomatic, AGE 180 degrees.

Left knee joint Asymptomatic; AGE 170 degrees.

Patient walked without a cane.

X-ray (4/12/54): *Left elbow. Better calcification of all osseous structures* and no additional soft tissue swelling.

Knee joints Some increase in calcium density but decalcification was still quite marked, less soft tissue swelling.

Examination (6/10/54): *Left knee joint.* AGE 160 degrees

Left elbow. Painless but motion was quite restricted with extension limited to 120 degrees.

Ambulatory Treatment (6/10/54 to 6/24/54): Manufacture and delivery of fiberglass appliance for extension of left elbow and a small fiberglass brace for left leg to be used for swimming.

Examination (11/22/54): *Left elbow:* AGE 170 degrees, almost asymptomatic

Left knee joint. AGE 170 degrees.

Right knee joint. AGE 180 degrees, AGF 150 degrees.

Examination (1/28/55). *Ambulation at home without braces; no recurrence* of pain or deformity

Right knee joint AGE 175 degrees, AGF 90 degrees Appearance and function so good that brace was no longer required

Left knee joint. AGE 160 degrees.

LHH Treatment (1/28/55 to 1/29/55, 1 day): For complete relaxation prior to making plaster model of left leg

X-ray (1/28/55): Knee joints· Symmetrical and satisfactory development of all osseous structures Appearance of right knee was most satisfactory with articular space very well preserved Left knee joint showed more marked decalcification than on right side There had been no further joint destruction

During this hospitalization, a plaster cast was made of left leg for a new hemophilia Hessing brace and patient was also measured for a Hewitt elastic appliance for right knee with posterior hinge and one-piece tongue

Ambulatory Treatment (2/19/55 to 2/26/55): New appliances were delivered and patient walked without discomfort.

Last Follow-up (2/21/56). Left knee joint Satisfactory condition except for continuing restriction of extension to 170 degrees

Right knee joint Asymptomatic.

Case #6495 (Age, 35)

History Married, three children, accountant (own business) No hemophilia in mother's family, 3 brothers, all hemophiliacs, 2 deceased.

Circumcision. Bleeding for 5 months Diagnosis not made at that time Many bruises when crawling and beginning to stand and walk

Before 5 years of age Both elbows deformed and calcified

Active school life with hemorrhages into right and left knees as well as internal bleeding.

1949 Extraction of teeth, bled for 6 weeks with 16 transfusions

1949-1953 Recurrent hemorrhages into right and left knee joints with severe pain and increasing disability

1950-1953 Left knee developed tendency toward locking, usually followed by hemorrhaging

5/8/53 Left knee locked in flexion When attempting to straighten the knee, an extremely painful hemorrhage occurred, with considerable muscle spasm and jerking. Bedridden for 5 days with ice bags on left knee

During the past year, patient had one transfusion for an intestinal hemorrhage. Until recently, there have been blood clots in the urine

First Orthopaedic Examination (6/1/53): Patient is crutch ambulatory with severe pain in left knee, cannot bear weight on left leg Sleep is greatly disturbed by this pain Right knee joint is not painful but right leg feels weak

Left knee Markedly painful, swollen, tender and hot Redness and considerable glossiness of skin on dorsum due to tension from recent hemorrhage AGE 130 degrees Inspection of original x-ray films dated 9/23/52 showed markedly diminished articular space Productive changes were most marked between femur and patella There were few juxta-articular cysts Shape of the patella was not characteristic of hemophilic arthropathy suggesting that most of bony changes originated after growth had ceased

X-ray (6/2/53). Knee joints Marked degree of hemophilic arthropathy Considerable soft tissue swelling at left knee in conjunction with the acute hemorrhage into knee joint

Elbows Mild hemophilic arthropathy with cystic hypertrophic changes

Ankles Mild hemophilic arthropathy with cystic hypertrophic changes

Feet Slight degree of generalized decalcification and slightly increased trabeculation

LHH Treatment (6/1/53 to 6/5/53, 4 days): To control acute and painful condition of left knee

After treatment with bed rest, ice, immobilization on posterior flexible knee splint, and transfusions with quick frozen plasma, the acute soreness of the left knee subsided Patient was discharged wearing two posterior flexible knee splints

Examination (7/28/53) Patient walked with left-sided limp Right knee was

almost completely extended and required no correction prior to fitting with hemophilia Hessing brace. Left knee joint was moderately enlarged, showing some tenderness on passive movements. AGE 140 degrees.

LHH Treatment (7/28/53 to 8/27/53, 30 days): For correction of flexion contracture of left knee

Flexion contracture of left knee responded to correction by Quengel and wedge casts Both legs were fitted with hemophilia Hessing braces with Lofstrand spring joints at the knee, ankle joints and sandals to be worn in regular oxfords The acute and painful condition of the left knee which followed repeated recent hemorrhages was finally relieved by Hydrocortone. Patient became ambulatory, bearing little weight on two crutches which he used largely for stability. He was permitted to drive a car.

LHH Treatment (4/7/54 to 4/9/54, 2 days): For extraction of abscessed tooth

As patient exhibited a psychotic reaction after a tooth extraction, he was transferred to Bellevue Hospital for observation for one week. Thereafter, he was returned to Lenox Hill Hospital for an additional two weeks of bed rest

Examination (5/4/55): Patient suffered great pain and recurrent superficial hemorrhages following a fall in January 1955 There had been no significant injury but pain in both knees was so great at times that Dilaudid® was required There was some discoloration of skin in region of the right knee and on both thighs and lower-legs from very superficial recent hemorrhages. No other significant changes in function of both knees since June 1953 was apparent. Patient walked without crutches

X-ray (5/4/55): *Knee joints* There was some degree of recalcification and better trabeculation of osseous structures since 6/2/53 and no evidence of a recent injury to the right knee joint Both knees again showed advanced destruction and deformity of all articular surfaces due to hemophilic arthropathy.

Ambulatory Treatment (5/4/55): Appliances required, Butazolidin® prescribed for pain in knee joints

Case #6311 (Age, 47)

History: Negative family history Married 21 years.

Since 11 years of age. Treated in Europe for what appeared to be tuberculosis Developed flexion contracture of right knee and right hip, and equinus deformity necessitating weight-bearing on ball of right foot only. Most of hemophilic involvement occurred during adult life.

Left elbow: Many hemorrhages into joint leading to juxta-articular bone destruction, pain and stiffness

Age 26: Became aware of hemophilia only after marriage.

Recent years: Patient attended a hemophilia clinic for weekly prophylactic injections These did not prevent recurrent hemorrhages into right knee, left elbow and, just prior to first orthopaedic examination, into right hip region.

Profession: Rotogravure pressman.

First Orthopaedic Examination (2/14/53): Patient walked with mild right-sided limp Suffered from extensive hematoma in right pelvitrochanteric region

with discoloration of skin extending through groin into scrotum. No clinical evidence of involvement of right hip joint proper. Musculature of right leg showed considerable atrophy.

Right knee joint. Slight deformity and restriction of motion from AGE 150 degrees to AGF 100 degrees. Considerable crepitation on passive motion.

Right foot: Dorsiflexion was restricted but no evidence of bone or joint involvement with hemophilic arthropathy.

Left elbow: Considerable enlargement, partly due to soft tissue swelling, partly to bone deformity from hemophilic arthropathy. Motion possible between AGE 130 degrees and AGF 60 degrees.

X ray (2/14/53): *Right side of pelvis and hip joint.* Right side of pelvis, particularly its distal half was smaller than identical structures on left side. There was considerable decalcification throughout right side of pelvis and right femur. Proximal end of right femur was smaller than left with considerable degree of coxa valga. The hip joint proper appeared normal.

Right knee joint. Lack of development of all osseous structures with moderate decalcification. There was thinning of articular cartilage and marked irregularity of articular surfaces. No evidence of juxta-articular cysts and no deformity of the patella as seen in most cases of hemophilia where involvement occurred early in life.

Left elbow. Quite marked and characteristic changes of hemophilic arthropathy with thinning of articular cartilage, enlargement of the distal extremity of humerus, juxta-articular cysts, most marked in ulna, and some valgus deformity of elbow.

Ambulatory Treatment (2/19/53 to 3/14/53): After plaster of Paris models were made, patient received a hemophilia Hessing brace for right leg with Lofstrand spring joints at knee, and a fiberglass appliance for the left elbow to be worn while at work.

Examination (10/25/55): *Left knee joint.* Since May 1955, patient had 6 hemorrhages into the joint which was not swollen but showed slight local heat. AGE 180 degrees, AGF 90 degrees. There was lateral instability with knee extended.

Right knee joint. No swelling, tenderness or local heat. AGE 170 degrees. AGF 70 degrees. Remained asymptomatic ever since hemophilia Hessing brace was used.

Patient no longer required hemophilia Hessing brace while working and wore it only occasionally at home. Had benefited greatly.

Ambulatory Treatment (10/27/55): Patient received Hewitt elastic brace with one-piece tongue for left knee.

March, 1956: Hospitalized for kidney hemorrhages, received 3 blood transfusions.

Last Follow-up (8/24/56): Letter reported no severe hemorrhages in past 2 years. Several hemorrhages into left elbow in past year were controlled by plasma.

Because of active nature of employment, patient had been wearing hemo-

almost completely extended and required no correction prior to fitting with hemophilia Hessing brace Left knee joint was moderately enlarged, showing some tenderness on passive movements AGE 140 degrees.

LHH Treatment (7/28/53 to 8/27/53, 30 days): For correction of flexion contracture of left knee

Flexion contracture of left knee responded to correction by Quengel and wedge casts. Both legs were fitted with hemophilia Hessing braces with Lofstrand spring joints at the knee, ankle joints and sandals to be worn in regular oxfords. The acute and painful condition of the left knee which followed repeated recent hemorrhages was finally relieved by Hydrocortone Patient became ambulatory, bearing little weight on two crutches which he used largely for stability. He was permitted to drive a car.

LHH Treatment (4/7/54 to 4/9/54, 2 days): For extraction of abscessed tooth

As patient exhibited a psychotic reaction after a tooth extraction, he was transferred to Bellevue Hospital for observation for one week. Thereafter, he was returned to Lenox Hill Hospital for an additional two weeks of bed rest.

Examination (5/4/55): Patient suffered great pain and recurrent superficial hemorrhages following a fall in January 1955. There had been no significant injury but pain in both knees was so great at times that Dilaudid® was required There was some discoloration of skin in region of the right knee and on both thighs and lower-legs from very superficial recent hemorrhages. No other significant changes in function of both knees since June 1953 was apparent Patient walked without crutches

X-ray (5/4/55): *Knee joints* There was some degree of recalcification and better trabeculation of osseous structures since 6/2/53 and no evidence of a recent injury to the right knee joint Both knees again showed advanced destruction and deformity of all articular surfaces due to hemophilic arthropathy.

Ambulatory Treatment (5/4/55): Appliances required, Butazolidin® prescribed for pain in knee joints

Case #6311 (Age, 47)

History: Negative family history Married 21 years.

Since 11 years of age. Treated in Europe for what appeared to be tuberculosis Developed flexion contracture of right knee and right hip, and equinus deformity necessitating weight-bearing on ball of right foot only. Most of hemophilic involvement occurred during adult life

Left elbow: Many hemorrhages into joint leading to juxta-articular bone destruction, pain and stiffness.

Age 26 Became aware of hemophilia only after marriage.

Recent years: Patient attended a hemophilia clinic for weekly prophylactic injections. These did not prevent recurrent hemorrhages into right knee, left elbow and, just prior to first orthopaedic examination, into right hip region

Profession. Rotogravure pressman.

First Orthopaedic Examination (2/14/53): Patient walked with mild right-sided limp. Suffered from extensive hematoma in right pelvitrochanteric region

with discoloration of skin extending through groin into scrotum No clinical evidence of involvement of right hip joint proper. Musculature of right leg showed considerable atrophy.

Right knee joint: Slight deformity and restriction of motion from AGE 150 degrees to AGF 100 degrees Considerable crepitation on passive motion.

Right foot. Dorsiflexion was restricted but no evidence of bone or joint involvement with hemophilic arthropathy

Left elbow: Considerable enlargement, partly due to soft tissue swelling, partly to bone deformity from hemophilic arthropathy Motion possible between AGE 130 degrees and AGF 60 degrees

X-ray (2/14/53): *Right side of pelvis and hip joint* Right side of pelvis, particularly its distal half was smaller than identical structures on left side There was considerable decalcification throughout right side of pelvis and right femur Proximal end of right femur was smaller than left with considerable degree of coxa valga. The hip joint proper appeared normal

Right knee joint Lack of development of all osseous structures with moderate decalcification There was thinning of articular cartilage and marked irregularity of articular surfaces. No evidence of juxta-articular cysts and no deformity of the patella as seen in most cases of hemophilia where involvement occurred early in life

Left elbow: Quite marked and characteristic changes of hemophilic arthropathy with thinning of articular cartilage, enlargement of the distal extremity of humerus, juxta-articular cysts, most marked in ulna, and some valgus deformity of elbow.

Ambulatory Treatment **(2/19/53 to 3/14/53):** After plaster of Paris models were made, patient received a hemophilia Hessing brace for right leg with Lofstrand spring joints at knee, and a fiberglass appliance for the left elbow to be worn while at work

Examination **(10/25/55):** *Left knee joint* Since May 1955, patient had 6 hemorrhages into the joint which was not swollen but showed slight local heat AGE 180 degrees, AGF 90 degrees There was lateral instability with knee extended

Right knee joint No swelling, tenderness or local heat AGE 170 degrees, AGF 70 degrees Remained asymptomatic ever since hemophilia Hessing brace was used

Patient no longer required hemophilia Hessing brace while working and wore it only occasionally at home Had benefited greatly

Ambulatory Treatment **(10/27/55).** Patient received Hewitt elastic brace with one-piece tongue for left knee

March, 1956: Hospitalized for kidney hemorrhages, received 3 blood transfusions

Last Follow-up **(8/24/56):** Letter reported no severe hemorrhages in past 2 years Several hemorrhages into left elbow in past year were controlled by plasma.

Because of active nature of employment, patient had been wearing hemo-

philia Hessing brace on right leg on alternate days, and Hewitt brace for left knee every day even during hot weather

Case #6516 (Age, 22)

History: No family history of hemophilia College student (teaching).
Age 18 months Diagnosis of hemophilia
Recurrent hemorrhages into elbows, right wrist, knees; ankles
Past five years· Ankles only.

March 1953 Admitted to Bronx Hospital for throat hemorrhages after coughing attack. Since then, recurrent hemorrhages into left ankle; severe pain on weight-bearing, wore Ace bandage, cane ambulatory.

First Orthopaedic Examination (6/11/53): Left ankle: Considerable enlargement *of joint and soft tissue swelling but no evidence of recent hematoma Small* subcutaneous, hard, movable nodule anterior to external malleolus and lateral to extensor tendons of toes may represent a calcium deposit or small loose body No significant restriction of motion No atrophy of musculature of left lower-leg Patient advised not to bear weight on this ankle without protection

X-ray (6/11/53). *Left ankle.* Definite involvement with hemophilic arthropathy Juxta-articular cysts, particularly in astragalus Some thinning of articular space, widening of articular surface of tibia and fibula, tilt of astragalus within mortise in direction of valgus, marginal lipping at all articular spaces Other osseous structures of tarsus, metatarsus and toes showed minimal degree of decalcification.

Right foot and ankle. Minimal degree of involvement of ankle joint. Articular space well preserved and no evidence of arthropathy

[illegible]

tails of skeleton

Examination (4/14/55). *Left foot and ankle:* Asymptomatic, no pain or swelling. Patient no longer required brace unless there was a recurrence of hemorrhaging

X-ray (4/14/55). *Left foot and ankle* Better calcification and better trabeculation, no evidence of progression of hemophilic arthropathy

Examination (5/4/55): Left ankle: Well localized pain and swelling along peroneus tendons throughout region of external malleolus Present disability due to tenosynovitis

Left foot In plano valgus position, had not been properly supported since *Hessing-type brace was discarded in March 1955*

Ambulatory Treatment (5/4/55): A plaster model was made for a Lange-type fiberglass foot plate and appliance was fitted and delivered

Case #5398 (Age, 23) (See Figure 13)

History· Known hemophiliac, one of three hemophiliac brothers, two maternal uncles died from hemophilia Not circumcised.

4 months. First hematoma

Hospital: 48 admissions, 260 transfusions

Age 12. Right shoulder joint affected, dislocated about 20 times since then, recently, anaesthesia necessary to reduce dislocations

Age 15 Left knee involved.

Age 19 Right hip injured in fall, became increasingly stiff Difficult ambulation, pain on weight-bearing

First Orthopaedic Examination (1/10/51): Right-sided limp, crutch ambulatory.

Right hip AGE 180 degrees, AGF 90 degrees, rigid in adduction of 15 degrees with internal rotation of 10 degrees Adduction contracture responsible for pelvic tilt down left side producing apparent shortening of right leg This was compensated for by a board under right foot of ¾ inch thickness

Left knee joint. Slight enlargement and moderate deformity due to productive arthritic changes and thickening of synovial membrane and capsule, most marked over median articular space where there was also maximum tenderness on palpation and pressure

Right shoulder: Atrophy of musculature and considerable restriction of motion Total range of motion was about one-third less than that on left side and there was crepitation on passive motion

X-ray (1/12/51): *Right hip* Pathologic changes suggestive of injury to neck or capital epiphysis of femur with secondary thinning of articular cartilage and some hypertrophic changes at margins of joint No involvement with hemophilic arthropathy

Right shoulder Involvement with hemophilic arthropathy Considerable decalcification of osseous structures

Left knee joint Involvement with hemophilic arthropathy Thinning most marked at internal articular space with spur formation at medial borders of tibia and femur

February 1951: Hemorrhage into left knee, hospitalized for 3 weeks

Examination (5/28/51): *Left knee joint* AGE 165 degrees, pain on weight-bearing Patient had considerable difficulty standing and walking Used cane General condition was poor

LHH Treatment (5/31/51 to 7/9/51, 39 days): For correction of deformities at right hip and left knee joint

Examination (5/31/51): *Right hip* Increasingly rigid

X ray (6/5/51). *Left knee joint* Considerable progress in involvement with hemophilic arthropathy and a greater degree of thinning of external articular space

Examination of right hip under Avertin anaesthesia showed a greater range of abduction and external rotation of right leg than anticipated Right leg was therefore immobilized in position of best possible abduction and neutral rotation at hip at the same time as a maintenance cast was applied to left leg Final appearance of cast was that of a double hip spica extending to above ankle on right side and including foot on left side A second cast was applied to the left

leg with knee in greater extension and AGE 175 degrees was obtained. A plaster of Paris model of the left leg was made for a hemophilia Hessing brace with Lofstrand spring knee joints. The brace was applied to the left leg with the knee held in complete extension Patient was discharged completely ambulatory with left knee free from pain and with an increased range of abduction at right hip.

Ambulatory Treatment (7/19/51): A model was made for a spiral-bar brace for right hip with Lofstrand spring joint at level of greater trochanter.

LHH Treatment (8/1/51 to 8/9/51, 8 days): Following contusion to left shoulder. Patient received unknown doses of Morphine and sedatives at home and at another hospital and was admitted in semi-comatose state.

Plasma administered, no evidence of involvement with hemophilic arthropathy, good recovery.

Case #6294 (Age, 25)

History: Mother's uncle died from hemophilia Not circumcised, diagnosis established soon after birth (blood tests). Repeated hospital admissions, more than 100 transfusions

Hemorrhages. Left knee, musculature of right forearm, kidney, left hip.

Age 12· Hemorrhages into left hip necessitating almost a year of bed rest during which time left knee developed flexion contracture. Left hip became and remained entirely asymptomatic.

Age 19. Hemorrhage into right forearm leaving hand with functional defect. Although originally right-handed, patient learned to use left hand.

Age 21: Started work as salesman. Excessive standing and walking produced hemorrhages into left knee with a permanent flexion contracture of varying degree

Age 25· Employed in an arcade where patient was required to stand for 8 hours This caused recurrent swelling and pain in left knee Patient observed an increase in deformity at left knee with a very painful area of bony prominence on medial femoral condyle

2/2/53 Transfusion with quick frozen plasma for painful swelling of left knee. Unable to work.

First Orthopaedic Examination (2/6/53): Fair general condition; left-sided limp, crutch ambulatory

Left knee joint. Enlarged and rigid in extension of 160 degrees; AGF 120 degrees Ballottement of patella indicated presence of blood within joint. There was moderate local heat, and maximum tenderness over a rough bony prominence of medial femoral condyle The tibia showed some posterior subluxation in relation to femur.

X-ray (2/6/53): *Left knee joint.* Differed from average hemophilic knee joint in several respects There was no deformity of patella; articular space was very

[illegible]

ably due to the fact that first hemorrhages into joint occurred when [illegible] was 21.

Ambulatory Treatment (2/6/53 to 3/5/53): After application of posterior flexible splint for immediate support of left knee, a plaster of Paris model was made of left leg for a hemophilia Hessing brace with Lofstrand spring joints at knee Condition of joint had improved to point where extension reached almost 180 degrees and swelling and ballottement of patella had disappeared Leg was again splinted until brace was delivered

Examination (5/13/53): *Left knee joint* Painless, slight ballottement of patella Patient reported that he worked regularly, wearing hemophilia Hessing brace constantly. Could not walk without it

Examination (9/16/53): *Left knee joint* New pain at external articular space and external femoral condyle. There was tenderness on palpation and pain on weight-bearing, particularly with knee fully extended

X-ray (9/16/53). *Left knee joint* Some improvement in calcium density of all osseous structures and some recalcification of large cystic area in internal femoral condyle. There was also an area of structural irregularity in anterior portion of articular surface of external femoral condyle which might have represented a new lesion responsible for patient's most recent complaint

Examination (4/15/54): *Left knee joint* Recurrent hemorrhages caused a return of some degree of flexion contracture Patient was hospitalized for several plasma transfusions Since hemophilia Hessing brace was repaired and worn continuously, most of contracture disappeared Well localized pain continued to exist over external articular space and proximal margin of external condyle of tibia

Examination (4/27/54): *Left knee joint* Less pain, more comfortable ambulation

Right hand Functional defect due to hemorrhage into musculature of right forearm 6½ years ago

X-ray (4/27/54): *Right hand* Minor involvement of 5th metacarpophalangeal joint

Ambulatory Treatment (4/27/54 to 5/18/54): Exercises to improve ulnar abduction at wrist and extension of fingers No orthopaedic appliances required

Examination (6/1/54): *Right hand* Unable to continue exercises for increased ulnar abduction at wrist because of pain and swelling at second metacarpophalangeal joint

Left knee joint Fully extended

Examination (7/6/54): *Left knee joint* Two hemorrhages into medial aspect for which 2 plasma transfusions were required Ace bandage to be worn on left knee when brace was discarded at night

9/14/54: Plasma given for mild hemorrhage into stomach

Examination (9/16/54): Patient was driving a car

Examination (1/18/55). Satisfactory gait.

X-ray (1/18/55): *Left knee joint* Further improvement in calcium density of all osseous structures Almost complete healing of large cystic area in internal femoral condyle.

3/3/55: Hematuria requiring plasma

Examination (4/28/55): *Left knee joint:* Patient complained of recurrent attacks of pain and swelling. Job required standing and walking for at least 8 hours per day. Examination revealed no significant swelling or irritation

Ambulatory Treatment (4/28/55 to 6/1/55): Patient was fitted with a Hewitt elastic knee brace to be used for support of the left knee while the left hemophilia Hessing brace was undergoing alterations, and for the summer.

Examination (9/9/55): During the 6 weeks' period that Hewitt elastic knee brace was worn, patient had occasional weight-bearing pain on medial aspect of left knee but no swelling or hemorrhaging. *He was instructed to wear the hemophilia Hessing brace only when the knee became painful or if he had to stand or lift more than usual.*

Examination (12/17/55): *Left knee joint. A slightly swollen and painful* bursa had formed over prominent lateral aspect of head of fibula. There was some tenderness and pressure over medial articular space but no local heat or swelling *Patient stated that he had to sway his body to the right to prevent pain on medial* aspect of the knee The joint was painless when he wore the hemophilia Hessing brace but he experienced some pain within the knee when the *Hewitt elastic knee brace was worn.*

Examination (3/17/56): *Right ankle:* During the 8 months prior to this examination, ankle joint was frequently swollen, with some pain on weight-
nternal malleolus from
here was some crepita-
dorsiflexion

Left knee· Asymptomatic

X-ray (3/17/56): *Right foot and ankle:* Some thinning of articular cartilage of joint and irregularities of articular surfaces of tibia and of astragalus, representing *slight involvement of joint with hemophilic arthropathy.*

Ambulatory Treatment (3/17/56 to 4/21/56): Patient was fitted with a strong elastic anklet This, however, was not sufficiently strong to prevent recurrent swelling and pain at ankle and patient was unable to work *He then received a* spiral-bar lower-leg brace for right foot and ankle Patient was advised to return to work after he had become accustomed to ambulation with the new brace.

Last Follow-up: 5/24/56.

Case #7389 (Age, 8) (See Figure 58)

History: Negative family history. Two sisters

Circumcision Bleeding could not be stopped by suture Patient was transferred to a hospital for transfusions

Ankles and knees Occasional *hemorrhages with temporary swelling*

November 1953 Serious involvement of left ankle followed by development of flexion contracture of left knee

Age 8 Hematuria, no other *internal hemorrhages.*

September and October 1954 Received two Hessing-type leg braces with Lofstrand spring joints at knees at Lenox Hill Hospital Brace Department on prescription of family physician

December 1954: Last hemorrhage into left knee

Complaints Unable to wear Hessing-type brace for left leg because of flexion contracture which developed after recent hemorrhage Hessing-type brace for right leg was still worn although right knee joint became swollen after wearing brace for several hours. Patient was crutch ambulatory bearing no weight on left leg.

First Orthopaedic Examination (1/12/55): Left knee joint: Moderately swollen and held in flexion of 115 degrees, considerable posterior subluxation of tibia Entire left leg showed marked atrophy.

Left ankle Swollen with some restriction of motion and crepitation indicating involvement with hemophilic arthropathy

Right knee joint: Appeared in much better condition than left, painless, no local heat. AGE 165 degrees Hessing-type brace did not fit well because spring joints extended to 180 degrees and knee could not bend beyond 165 degrees

X-rays (Inspection 1/12/55 of films dated 8/16/54): *Right knee joint* AGE 165 degrees Articular space was fairly well preserved but all articular surfaces, including patella, showed involvement with hemophilic arthropathy including juxta-articular cysts throughout the distal femoral epiphysis particularly on medial side. Transverse diameter of femoral condyles was definitely enlarged. There was a moderate degree of generalized decalcification, some posterior subluxation of the tibia in relation to the femur and increased soft tissue density throughout joint

Left knee joint There was a greater degree of generalized decalcification and more posterior subluxation of the tibia than on the right side On the whole, the degree of hemophilic arthropathy was more marked at the left knee although the right knee presented a fully developed picture.

Left ankle and foot Moderate degree of generalized decalcification of all osseous structures and considerable destruction of ankle joint proper due to deformity of articular surfaces of astragalus and tibia

Right foot and ankle No significant involvement with hemophilic arthropathy

Diagnosis: Knees and left ankle Hemophilic arthropathy of considerable degree, flexion contracture of 115 degrees at left knee

LHH Treatment (1/17/55 to 1/29/55, 12 days). For hemophilic arthropathy of knees and left ankle, and flexion contracture at left knee

X-ray (1/17/55): *Left knee joint* All osseous structures showed marked decalcification Articular surfaces of femur and tibia showed marked involvement with hemophilic arthropathy Patella was smaller than on right side and was squared off at the distal pole. There was some posterior displacement of tibia in relation to femur and quite marked genu valgum deformity.

Right knee joint AGE 165 degrees Involvement with hemophilic arthropathy was much less marked than at left knee.

Right ankle: Slight degree of irregularity of articular surfaces and some soft tissue swelling around joint

Left ankle Definite involvement with hemophilic arthropathy, most marked at articular proximal surface of astragalus and ankle joint

During hospitalization, correction from ACE 115 degrees to ACE 150 degrees at left knee was obtained by means of a Quengel cast hip spica with subluxation hinges at knee. Patient continued to wear *his Hessing-type brace on* right leg to maintain extension at 165 degrees. After correction of flexion contracture at left knee, patient was able to wear the old Hessing-type brace with knee joint locked at 150 degrees. A well-padded leather knee cap was added. Patient was also treated for an upper respiratory infection during this admission

Ambulatory Treatment (2/5/55 to 5/2/55): The lower-leg sections of the braces were lengthened Patient attended school using crutches. Left knee remained asymptomatic. Spring tension at left knee was increased

LHH Treatment (5/2/55 to 5/3/55, 1 day): For acute hemorrhage into left foot and ankle since 4/29/55 and flexion contracture of 120 degrees at left knee.

Examination (5/2/55). *Left foot and ankle:* Moderate swelling with some discoloration of skin, minimal degree of local heat and some restriction of motion.

During overnight admission, a protective cast was applied to entire left leg and prepared for later wedging at knee

Examination (5/12/55): *Left ankle.* Swelling, pain and restriction of motion had disappeared

Left knee ACE 140 degrees with rather marked posterior subluxation of tibia

Ambulatory Treatment (5/12/55 to 6/28/55): Cast, applied to left leg from toes to below groin, was wedged twice at the knee, increasing extension there to 155 degrees Hessing-type brace was refitted and patient was fitted with a Hewitt-type elastic knee brace without posterior hinge for right leg, to be worn occasionally during summer in place of the Hessing-type brace. Patient still preferred to be crutch ambulatory.

Examination (9/22/55): *Right knee joint·* Patient was unable to wear Hewitt elastic knee brace during summer because knee became swollen and painful as soon as Hessing-type brace was discarded. ACE 165 degrees. An attempt to further extend knee produced muscle spasm. Advisable to lock Lofstrand spring joints at knee in position of best possible extension until musculature was relaxed

Left knee joint ACE 155 degrees

Ambulatory without crutches.

LHH Treatment (11/1/55 to 11/4/55, 3 days): For correction of residual flexion contractures at right and left knees.

X-ray (11/1/55): *Knees.* Some degree of recalcification and no soft tissue swelling Involvement of articular surfaces with hemophilic arthropathy showed some progression since 1/17/55 Posterior subluxation of tibia in relation to axis of femur was noted bilaterally

During this admission, models were taken for hemophilia Hessing braces and a double hip spica cast was applied extending from lower ribs to toes and wedged at the knees.

Ambulatory Treatment (11/10/55 to 12/26/55): Cast was again wedged at knees When cast was removed, extension at right knee was limited to ACE 165 degrees, at left knee to ACE 160 degrees with persistent posterior subluxation of tibia in relation to femur. New hemophilia Hessing braces were delivered

Last Follow-up (5/22/56, patient still under treatment): Plaster of Paris models were made for new hemophilia Hessing braces for both legs

Note: No further progress with the left leg had been made because of the unusual degree of posterior subluxation of tibia that existed For this reason, construction of a special knee joint, to be the first of its kind, was required for the new hemophilia Hessing brace

Case #6004 (Age, 14)

History. One sister Hemophilia family tree great uncle, cousin, second cousin (dead)

Age 1½. After measles, patient developed tumor at neck Surgery was accompanied by extensive hemorrhages and patient barely survived

Age 9 Right knee involved

Age 13. Left knee suffered first hemorrhage after trauma

Elbows· Slightly affected after injuries, left elbow showed residual stiffness

Internal One episode of hematuria.

No evidence of cycle, all hemorrhages occurred as result of trauma

Past treatment Several whole blood transfusions once or twice a year

Able to attend school regularly, active swimmer, could drive boat and car

First Orthopaedic Examination (6/8/52): Well developed boy in good general condition Could bear weight without crutches but stance was awkward due to marked flexion contractures of knees

Right knee joint Typical appearance of moderate hemophilic arthropathy (more marked than on left side) with muscular atrophy AGE 110 degrees, AGF 100 degrees

Left knee joint Range of motion from AGE 120 degrees to AGF 90 degrees, at times, knee could be almost completely extended Moderate hemophilic arthropathy.

LIHH Treatment (6/18/52 to 7/26/52, 38 days). For flexion contracture at knees

During hospitalization, flexion contractures of knees were almost completely corrected by Quengel and maintenance casts for right knee and wedge cast for the left Patient was fitted with hemophilia Hessing braces with Lofstrand spring joints at knees made to plaster models and was discharged able to stand and walk using a walker He also received two posterior flexible knee splints to be worn for swimming.

Examination (9/16/52): *Knees·* Painless, maintenance of almost complete extension

Left hip: Flexion contracture, resulting from hemorrhage, extension limited to 155 degrees

X-ray (9/16/52): *Left hip* No bone or joint involvement.

Ambulatory Treatment (9/16/52 to 9/17/52): Flexion contracture at left hip corrected To avoid development of flexion contractures, patient was advised to have plasma transfusions at first indication of pain or hematoma in this region

Examination (8/25/53): Excellent progress, no recurrence of hemorrhages

into knees, ambulated without cane and with knees rigid in extension; two episodes of internal hemorrhages.

Left knee joint AGE 180 degrees with slight degree of valgus, less marked when standing than when supine.

Right knee joint: AGE 170 degrees with posterior subluxation of tibia.

Both knees had only a few degrees of flexion.

X-ray (8/25/53): *Pelvis and hip joints:* No evidence of hemophilic arthropathy.

Knees. Generalized decalcification of osseous structures and moderate deformity of femoral and tibial condyles and their articular surfaces due to involvement with hemophilic arthropathy. Destruction of articular space and changes at patella were somewhat more marked at left knee Joint was held in complete extension with correct lateral alignment of femur and tibia. Clinical impression of valgus deformity was not substantiated by x-ray evidence. Articular space was markedly diminished at both knee joints. At right knee, there was a considerable degree of posterior dislocation of tibia in relation to long axis of femur This joint showed slightly better preservation of articular space than on left side.

Ambulatory Treatment (8/25/53 *to* 9/1/53): Plaster models were made of both legs for new hemophilia Hessing braces with Lofstrand spring joints and simple milled ankle joints and sandals. Appliances were delivered and patient's gait was better than ever.

Examination (12/14/54): Patient had been walking at home for short periods without braces.

Knees: Painless, free from hemorrhages, completely extended, with continued deformity on right side due to posterior subluxation of tibia A few degrees of flexion were possible at left knee while right knee appeared rigid in extension

X-ray (12/14/54): *Knee joints* Considerable recalcification of all osseous structures Articular space at right knee was not as well preserved as it had been in 1953, although there was no evidence of progress of hemophilic arthropathy

Ambulatory Treatment (12/14/54 *to* 12/21/54): Patient was advised to test stability of knee joints by walking for a day or two without appliances. Impression was that motion might be encouraged at left knee while right knee might become ankylosed in complete extension. Clinical and x-ray findings suggested that hemophilia Hessing braces were no longer necessary except for emergency use Plaster models of legs were taken and old braces fitted with new thigh sections and generally repaired Patient received Hewitt elastic knee braces with posterior hinge and one-piece tongue and a pair of Whitman foot plates He reported that a good deal of ambulation without braces produced no pain or discomfort at either knee.

Last Follow-up (8/23/56): A letter from the patient's father reported that for long periods after his last visit to New York City for orthopaedic treatment, the boy was quite well, missing school only occasionally and remaining with his high school class. He was planning to attend college During one period in the

winter of 1955 and another in the summer of that same year, he suffered hemorrhages into joints and muscles around a hip, and required several plasma transfusions During his senior year in high school, 1955-1956, he lost no time in the fall, but after Christmas he had another hemorrhage into the hip joint with recurrences in July and August 1956, resulting in a stiffening of the hip The parents observed no specific trauma to which these hemorrhages could be traced but felt that bleeding occurred when the patient became overtired. He had no further orthopaedic treatment since December 1954 and was completely ambulatory without appliances except during periods of illness and convalescence.

Case #6140 (Age, 6½)

History: Prolonged bleeding at circumcision Diagnosis of hemophilia was not established at that time. This was followed by appearance of many bruises, prolonged bleeding after injury to tongue, and recurrent hemarthros in knees and ankles These hemorrhages into joints were soon absorbed and joints functioned normally No internal hemorrhages

Age 2-4 years Frequent examinations by hematologist Diagnosis of pseudohemophilia with prolonged bleeding time and clotting time attributable to abnormal fragility of capillaries

July 1952 When patient was 6 years of age, he suffered an injury to right ankle which became painful and swollen X-ray, taken on 7/8/52, led to diagnosis of chip fracture from internal malleolus Cast was applied and removed after one week Patient could not bear weight on ankle which remained painful Another set of x-rays was taken and diagnosis of chip fracture confirmed by orthopaedic surgeon Heavy walking cast was applied and worn for three weeks After it was removed, patient was still unable to bear weight on right foot although there was no pain A marked equinus deformity developed Patient recently received two transfusions of quick frozen group specific plasma without success Unable to return to school as he could not walk properly

First Orthopaedic Examination (10/14/52): Well developed, nervous child, limp, crutch ambulatory

Skin. Numerous but small areas of discoloration from recent bruises

Right leg Musculature showed definite but moderate atrophy in comparison with left side Patellar and Achilis reflexes were active and equal

Right ankle Fixed equinus position of 160 degrees Joint showed slight swelling, particularly to right and left of tendo Achilis

Right knee joint No involvement with hemophilic arthropathy, functioned well

Left knee joint No involvement with hemophilic arthropathy

Left ankle No involvement with hemophilic arthropathy

Inspection of series of x-ray films taken between 6/26/50 and 10/2/52 showed soft tissue swelling and increased shadow density at ankles and knees but no evidence of hemophilic arthropathy or permanent thickening of capsules

Inspection of films of right ankle taken after injury to joint on 7/8/52, showed small, irregular shadow of calcium density in close proximity, distal to tip of

internal malleolus. There was no defect in contour of distal epiphysis of tibia The shadow, which was interpreted as a fracture from the internal malleolus, in all probability represented calcium deposits from previous hematoma in this area Films of right ankle, taken on 10/2/52, when so-called fracture was pronounced healed, showed same crescent-shaped shadow distal to tip of internal malleolus. At that time, the shadow was slightly smaller and denser. Most recent films of right ankle suggested possibility of slight irregularity and squaring off of articular surface of astragalus.

Discussion Disability exhibited at first orthopaedic examination resulted from marked and fixed equinus position of right foot which was caused neither by previous hemorrhages into soft tissue structures of right calf nor by bone deformity at right ankle, but in all probability by immobilization in plaster cast for an alleged fracture of the internal malleolus.

LHH Treatment (10/14/52 to 10/26/52, 12 days): *X-ray (10/15/52): Ankles*· Fixed equinus of right foot with ankle at 155 degrees Generalized decalcification of all osseous structures with some rather dense calcification near tip of internal malleolus Articular surface of astragalus irregular with destruction of cartilage and some of the juxta-articular bone. Contour of articular surfaces showed definite squaring off or flattening Similar changes noted at left astragalus although there were no clinical symptoms concerning left foot.

During hospitalization, a fixed equinus position of right foot with an angle of 155 degrees and with x-ray evidence of early signs of hemophilic arthropathy, was corrected by Quengel cast applied to right foot and lower-leg incorporating aluminum bars on sole of foot and below knee Correction was made to an angle of 90 degrees A maintenance cast was then applied for walking

Ambulatory Treatment (11/4/52 to 11/18/52): Patient walked very little. When bearing weight on right leg, he held knee in flexion and most of weight was borne on ball of foot As it was not advisable to continue with this type of gait which placed too much strain on right knee joint, a felt wedge was placed in heel of shoe so that patient could bear full weight on heel of cast and walk with knee extended After a plaster model was made of right lower-leg, a second walking cast was applied. Patient received a molded leather jointless brace for lower-leg and foot, holding ankle at 90 degrees. Within a week, he was able to walk extremely well

Examination (1/20/53): Non-ambulatory for 2 weeks. When appliance was received, patient walked well and appeared anxious to return to school but then complained of pain Very active at home, going up and down stairs but mostly on hands and knees Ambulation only with right leg in marked external rotation Bedridden 11 days with tonsillitis infection, refused to walk thereafter

Examination ruled out pain on weight-bearing. Patient walked with right-sided limp and tendency toward external rotation of right leg, due partly to fact that immobilization of right ankle necessitated walking with stiff knee held in extension which in turn made right leg too long in comparison with left Heel of right shoe to be decreased one lift while one lift should be added to left heel

Patient encouraged to walk Most of disability on walking was of psychogenic origin

3/2/53. Frequent tonsillitis since 1/20/53 Had not walked since

Examination **(3/9/53):** No weight-bearing on right leg Did not admit to pain at right foot or ankle Right-sided limp threw weight onto left leg Held *entire right leg stiff in complete extension*

Right foot and ankle No pressure marks after removal of brace, no significant swelling and no evidence of local heat or tenderness on pressure While in brace, foot had been kept at 90 degrees Active and passive motion was possible to 75 degrees No attempt was made to increase plantar flexion beyond 110 degrees. There was some crepitation on passive motion originating from the ankle joint proper Inasmuch as right foot had been rigid in equinus position since injury in July 1952, correction of equinus deformity since 10/16/52 was most satisfactory Determination to be made of degree to which osseous structures showed decalcification and involvement with hemophilic arthropathy It was possible that decalcification due to disuse might cause pain on weight-bearing There was surely an organic nucleus for patient's complaints but psychogenic element seemed to be in the foreground

X-ray (3/9/53): *Right foot and ankle* No significant recalcification of osseous structures since 10/15/52 Equinus position of 155 degrees had been completely corrected. Destruction of articular cartilage and some of juxta-articular bone of the astragalus and corresponding changes in articular surface of tibia, were of about same degree as on 10/15/52, although there was slightly more thinning of articular cartilage of ankle joint proper. Films showed organic basis of patient's complaints of pain on weight-bearing when ankle joint was not completely immobilized and foot well supported Recalcification possible only by "protected weight-bearing"

Ambulatory Treatment **(3/11/53 to 4/4/53):** A well padded walking cast was applied to right leg from toes to below knee with ankle held at 90 degrees Heel and instep, most sensitive throughout treatment since 10/15/52, were carefully protected with foam rubber.

A simple rubber lift was fitted with a small plate and leather straps to be used like a removable Stryker cast heel With this heel in place, patient was able to bear *weight and become ambulatory without pain Walked better without* cane but had to concentrate on keeping knee completely extended and bearing all weight on cast heel Patient walked very little, complaining of pain at heel when bearing weight Had not learned to walk with stiff right leg After removal of cast, appearance of right foot and ankle was satisfactory Considerable range of active motion was present with dorsiflexion possible to at least 80 degrees

X-ray (4/14/53). *Right foot and ankle* Involvement of right ankle with hemophilic arthropathy of moderate degree was sufficient to explain weight-bearing pain in spite of immobilization in a cast No change in appearance since last films were taken.

Ambulatory Treatment (4/14/53 to 6/2/53): There was no indication for another cast Patient required a Hessing-type lower-leg brace with ankle joint and sandal, built to restrict ankle motion to ACE 110 degrees, AGF 80 degrees Brace to transmit considerable part of weight-bearing to lower-leg, particularly to tuberosity of tibia. This would unweight the ankle joint proper to some extent. Appliance was delivered on 4/23/53. By 6/2/53, patient still held right leg stiff and walked with limp, using one cane.

X-ray (6/24/53): *Right foot and ankle* Minimal increase in irregularities of articular surfaces of ankle joint, foot and ankle showed some degree of recalcification of all osseous structures.

Examination (6/27/53): Patient still walked with right-sided limp, using cane, and performed some awkward movements which were partly psychogenic Definite tendency toward recurrence of equinus, or at least toward restriction of dorsiflexion beyond 100 degrees, forcing patient to walk with stiff and completely extended knee.

Right leg Musculature showed considerable atrophy.

Ambulatory Treatment (6/30/53 to 7/2/53): Patient received small plastic night splint to counteract tendency toward recurrence of equinus deformity. This was made to hold the right foot at an angle of 90 degrees.

Examination (11/11/53): Splint was not used; correction of equinus was so good as to render it unnecessary.

X-ray (11/11/53): *Right foot and ankle:* Improved calcium density in osseous structures, slight increase in irregularities of articular surfaces.

Examination (3/2/54): Patient walked well without cane.

Right foot and ankle No pain and no swelling of soft tissues. Ankle held rigid at 90 degrees without valgus tendency.

Right calf. Musculature of better quality.

Left foot In February 1954, patient suffered hemorrhage into left ankle. Treated with cast, ice and one Hydrocortone injection. Pain and swelling subsided rapidly Foot showed moderate valgus position. There was some soft tissue swelling between tendo Achillis and internal malleolus. Joint proper appeared free.

Ambulatory Treatment (3/2/54): Patient to wear nylon elastic stocking on left as well as on right leg to prevent swelling of ankles

X-ray (4/15/54): *Left ankle* Slight irregularities of articular surfaces of joint suggested that recurrent swelling in region of internal malleolus might be due to joint involvement rather than to repeated extrinsic trauma

Examination (7/2/54): *Left foot.* Marked valgus position requiring correction

Ambulatory Treatment (7/2/54 to 7/13/54): Patient received Lange-type fiberglass plate for right foot and Whitman-type fiberglass plate for left foot Hessing-type brace for right lower-leg to be discarded. Patient permitted to swim

Ambulatory Treatment (1/27/55 to 2/5/55): Patient received new and strong Whitman-type plate for left foot to correct excessive valgus position.

7/7/55. Hemorrhages into abdominal wall, right groin and into back or chest.

Hospital Treatment (not LHH) (7/7/55 to 7/15/55): Codeine, wet compresses, antihemophilic plasma. Hemorrhages probably were caused by sprain during swimming. No involvement of bone or joints

Examination (8/29/55): *Right foot and ankle* No swelling or deformity. Well supported by Lange-type foot plate

Left foot Asymptomatic but in plano valgus position of highest degree Whitman-type plate corrected as well as could be expetced and was well tolerated

Patient required no orthopaedic treatment, attended school

X-ray (4/17/56): No further articular or skeletal changes

Case #6028 (Age, 26)

History Mother's brother died at age 6 from blood condition, presumably hemophilia One brother, free of hemophilia

Before age 3 Diagnosis followed considerable bruising Confirmed when patient cut his tongue and required three transfusions to stop hemorrhaging

Age 3-14 years: Nosebleeds and joint bleeding were chief problem

Age 14 to 15 years Bedridden because of recurrent hemorrhages

Cycles Major cycle in spring, minor cycle in fall Bleeding occurred, however, at all times

Cycle sequence First, bleeding into joints, second, subcutaneous and intramuscular, third, from kidneys Most bleeding resulted from infection, usually associated with sore throats, and from lowered resistance

Transfusion history Age 3-8, Direct, Age 8-21, Whole blood, Age 21-23, Fraction I Red Cross, Age 25-26, Cutter's dried plasma

April 1951: Left knee suffered serious hemorrhage After aspiration, it never returned to normal as all joints had done in past after hemorrhages Knee remained weak and appeared to be regaining strength very slowly Ambulation was accompanied by pain and fatigue

May 1951-February 1952 Hemorrhages into left elbow averaged one every 2 weeks Arm could not be used Plasma introduced at first sign of hemorrhage (within 2 hours) stopped bleeding immediately, whereas if patient waited until second day after swelling developed, plasma was of little or no help in relieving swelling

Using counter pressure against articular and intramuscular hemorrhaging, patient was able to minimize area of swelling Rutin (300-400 mg per day) for four months was used with encouraging results and there were no major hemorrhages during this period

Present complaints Pain and disability originating from left elbow and left knee Patient wore no orthopaedic appliance.

First Orthopaedic Examination (6/28/52): Patient was cane ambulatory, walking with rather stiff legs and with a slight left-sided limp

Left knee joint Moderate swelling with tenderness over internal articular space Patella appeared plump AGE 180 degrees with some lateral instability apparently originating from internal lateral ligament, AGF 90 degrees Marked crepitation present on passive movements.

Right knee joint. Appeared almost normal with some crepitation on passive motion.

Left elbow Moderately enlarged with considerable restriction of motion AGE 140 degrees, AGF 80 degrees

X-ray (6/28/52): *Left elbow.* Increased soft tissue density on ulnar and posterior aspect of joint and some deformity of head of radius. Involvement with hemophilic arthropathy was most marked throughout ulnar part of joint Films suggested some cystic changes in proximal shaft of ulna. Articular space showed marked diminution.

Left knee Considerable decalcification of osseous structures in comparison with right Medial articular space was narrow suggesting thinning of medial meniscus. Patella was larger than that of right knee and showed in lateral projection increased A-P diameter but it had not lost its tip There was irregular soft tissue density Articular surfaces of tibial and femoral condyles suffered most on medial side. There were some productive changes at tibial spines and some widening of femoral condyles. Films showed no evidence of juxta-articular cysts

Right knee Better calcium density and structural details. Patella lay close to femoral condyles and was plump but not as large as left patella Medial articular space showed considerable thinning with irregularities of both femoral and tibial articular surfaces. The right knee joint was better preserved than the left and revealed no characteristic picture of hemophilic arthropathy.

Ambulatory Treatment (6/28/52): Since patient could not remain in New York for further investigation and treatment, he was advised to wear a Hewitt-type elastic knee brace for protection and stabilization of left knee. This was fitted with a special spring to improve extension of knee joint

Examination (11/19/52): Several hemorrhages into right knee and left elbow Hewitt brace for left knee was entirely successful in keeping joint completely extended and was probably responsible for protecting the left knee from the hemorrhages which afflicted other parts of the body Patient had constructed a crude celluloid splint for left elbow holding it in flexion of 90 degrees

Ambulatory Treatment (11/19/52 to 11/25/52): Received two new orthopaedic appliances for right knee, a Hewitt-type elastic brace, for the left elbow, a light-weight Hessing-type brace with elbow joint. Spring attachment was removed from Hewitt brace for left knee.

Examination (5/21/53): Patient secured a brace for the right elbow similar to that provided at LHH for the left. Complained of muscular weakness, particularly of left thigh and both arms. Recurrent hemorrhages into right elbow and into extensor muscle group of right forearm.

Suggested treatment Heavy resistance exercises that did not risk hemorrhages to improve muscle power of extremities.

Mail Follow-up (8/22/56): Patient reported that he had not been well but that conditon was under control. Since last orthopaedic examination in New York 3½ years prior to this communication, patient had 90 transfusions of frozen plasma administered ambulatory, all of which had been for hemorrhages into elbows and knees. Because plasma was given at first sign of bleeding into joints,

hospitalization had not been necessary After transfusions, patient followed regime of semi-immobilization, elevation and refrigeration for 24 to 48 hours Follow-up transfusions were given about one-third of time for recurrence of hemorrhages, usually 48 hours after the first transfusion

There had been no internal or muscular hemorrhages in 5 years Orthopaedic treatment had consisted largely of physical therapy Treatment was begun on March 2, 1955 and included gentle exercises for knees and elbows in a Hubbard tank at a temperature of 100°F Patient was getting in and out of tank by means of stretcher Report from the physiotherapy department at the out-of-town hospital where he was receiving treatment described the patient and his treatment as follows: "Limitation of movements in knees, ankles and elbows, sharp pains and dull aches in joints and muscles, general weakness of muscles The exercises were progressed slowly and the patient began to get in and out of the tank without a stretcher about 6 months later The condition of joints and muscles improved very slowly and the intervals between attacks of sharp pain began to be longer In July 1955 resisted exercises with springs were added to the free exercises in the Hubbard Tank." During the entire period, patient was undergoing x-ray therapy At date of the report from the hospital, June 12, 1956, range of movement in both knees was good. There was some limitation of extremes of flexion and extension

Patient believed that since he had been exposed to x-ray therapy, there had been a marked reduction in joint pain and tension This was most beneficial, he felt, as pain between hemorrhages, especially in elbows, was a serious problem Patient was working and ambulatory without appliances He believed that he should use a cane but his elbows could not tolerate this

Case #5653 (Age, 6)

History: No evidence of hemophilia in family or ancestry.

Clinical onset Infancy

Hospitalized at an early age for treatment of hemophilia and hemophilic arthropathy of left knee with flexion contracture of 90 degrees Subjected to aspiration and injection of Wydase®, traction, application of wedge casts After patient became ambulatory, last cast was removed on 8/10/51 with knee extended to 150 degrees. Severe pain, patient could not walk

First Orthopaedic Examination (8/11/51): Patient was bedridden, appeared to be extremely active and alert, parents reported behavior difficulties

Left knee joint AGE 140 degrees, pain, swelling contour not well defined

X-ray (8/11/51): *Left knee joint* Some decalcification of osseous structures and increased shadow density throughout joint Articular space was well preserved. No definite evidence of bone pathology

LIHI Treatment (8/11/51 to 8/18/51, 7 days): A cast was applied from toes to below groin to immobilize and protect left leg After second cast was applied, AGE 175 degrees was obtained Pain and swelling disappeared, and patient was discharged ambulatory, bearing full weight on left leg

Ambulatory Treatment (8/28/51 to 9/6/51): A model was made of the left

leg for a molded leather jointless brace holding leg in complete extension. A maintenance cast was then applied until brace was delivered.

Examination (9/8/51): *Left ankle:* Some swelling to both sides of tendo Achillis as well as around external malleolus. Foot held in moderate supination contracture of unknown etiology. Patient had not walked since joint became painful on 8/29/51. No weight to be borne on left foot until free range of motion was regained at ankle.

Examination (9/26/51): *Left foot and ankle:* No swelling or residual supination contracture. Slight stiffness in all directions.

Schooling: To attend first grade, regular school

LHH Treatment (10/13/51 to 10/23/51, 10 days): For hemorrhage into left knee joint. Plasma was administered and Alidase® injected into left knee joint. Following another hemorrhage into musculature of left calf, flexion at left knee joint was increased to AGF 90 degrees After transfusions and refrigeration, a maintenance cast was applied to left leg.

Ambulatory Treatment (11/1/51 to 11/15/51): When cast was removed, left knee joint showed a range of motion from AGE 130 degrees to AGF 90 degrees Application of two additional casts increased extension to AGE 165 degrees The old molded leather-steel brace was reapplied and patient was permitted to be ambulatory, bearing full weight on left leg, and to return to school.

Examination (12/1/51): *Left knee joint* AGE 180 degrees, with 20 degrees of active flexion.

Examination (7/3/52): Patient had been swimming using only splint and Ace bandage and was ready to discard appliances at beach

X-ray (7/3/52): *Left knee joint.* Very few changes Articular space was well preserved and there was hardly any decalcification.

Examination (9/27/52): Difficult education problem, extremely wild, constantly suffering new bruises and hemorrhages Required assistance of social worker.

Ambulatory Treatment (9/27/52 to 11/15/52): As left knee joint had developed good stability and motion without recurrent hemorrhages, a Hewitt elastic knee brace with one-piece tongue was indicated Pedes plano valgi II/III required correction with Whitman-type foot plates These appliances were fitted and delivered.

LHH Treatment (3/12/53 to 3/17/53, 5 days): For hemorrhage into left ankle.

Examination (3/12/53): *Left ankle:* Swelling, pain; foot held in equinus position with no motion.

A protective cast was applied to the left leg Patient made an almost complete recovery and cast was removed.

X-ray (after removal of cast): *Left ankle:* Soft tissue swelling and effusion in ankle joint Some generalized decalcification of all osseous structures. No evidence of involvement with hemophilic arthropathy.

April 1953 to June 1953: Small hemorrhages in right ankle and left knee.

Examination (9/24/53): *Knees* Fairly movable.

Patient had discarded all orthopaedic appliances during summer with resultant hemorrhages into ankles.

Examination (4/20/54): Left knee joint: Pain, swelling, AGE 150 degrees, as result of injury on 3/29/54

LIHI Treatment (10/26/54 to 11/6/54, 11 days): For acute hemarthros with flexion contracture at left knee.

Examination (10/26/54): *Left knee joint* Marked swelling and pain with ballottement of patella and local heat AGE 140 degrees

Hemorrhage into left knee subsided after transfusion with quick frozen group specific plasma given on 10/26/54 before admission to the hospital This was followed by complete immobilization of left leg in protective cast The flexion contracture was corrected to AGE 160 degrees and this position maintained by means of a cylinder cast permitting ambulation.

During this hospital admission, patient was also treated for marked anemia with whole blood transfusion, and with antibiotics for a mild upper respiratory infection

Ambulatory Treatment (11/30/54 to 12/7/54): Fitting and delivery of new hemophilia Hessing brace for left leg

12/7/54 Hemorrhage into right elbow.

March 1955. Hemorrhages into right leg Bedridden for two weeks Hemorrhage into left ankle during this period

Examination (3/29/55): Left leg had grown much longer than right, causing pelvic obliquity when patient was standing. He wore ½ inch lift on heel of right shoe to compensate for brace but required ¾ inch more The left knee joint was freely movable between AGE 175 degrees and AGF 70 degrees

Examination (12/10/55): Had suffered hemorrhages into left ankle and foot and hemophilia Hessing brace was discarded at that time Whitman foot plate was also discarded as it had become too small Hemorrhages also occurred into right ankle and elbows.

Left knee joint Hardly any restriction of complete extension, no swelling, local heat or tenderness

Left ankle Slight swelling with local heat at site of swelling Most soft tissue thickening was around external malleolus Dorsiflexion limited to 95 degrees

Elbows Restriction of complete extension by about 5 degrees on both sides, and of supination by not more than 10 degrees at left forearm

Right ankle: No abnormal findings

Plaster casts to be applied to elbows at home to protect and immobilize

Ambulatory Treatment (July 1956): Manufacture and delivery of two spiral-bar leg braces.

Last Follow-up (9/30/56): Letter reported that as patient was absent from school 50 per cent of time during the year, he required home instruction New braces were well tolerated

Case #6131 (Age, 4½)

History: No family history of hemophilia Patient's two brothers, ages 8 and 3, had no hemophilic symptoms

Circumcision: No abnormal bleeding

6 months: First hemorrhage into right knee

Age 1 year. Hospitalized three times for cuts.

No internal hemorrhages.

Series of hemorrhages into right and occasionally into left knee. Patient was always incapacitated for a short time but in the past had made a complete recovery Extremely active child

June 1952. Hemorrhage into right knee which remained painful and swollen Patient was unable to extend knee

X-rays to date: Reported negative and diagnosis of hemophilia and hemophilic arthropathy was not definitely established.

Present disability. Unable to walk

First Orthopaedic Examination (10/9/52): Right leg. Moderate muscular atrophy, most marked at thigh

Right knee joint Enlarged with local heat and restriction of motion from AGE 110 degrees to AGF 60 degrees

Left knee joint. Enlarged, AGE 170 degrees, AGF 50 degrees.

Pedes plano valgi II

Diagnosis Hemophilia, considerable hemophilic arthropathy of right knee; mild hemophilic arthropathy of left knee.

Hematological examination: Hemophilia diagnosis definitely established

X-ray (10/9/52): Right knee joint Small juxta-articular cysts and large marginal defect on lateral femoral condyle as well as thinning of articular space and irregularity of articular surface. Patella showed an early stage of the usual deformity seen in hemophilia It had grown faster than the left patella and was almost one-third larger

Left knee joint Bony structures showed only minimal changes of articular surface while joint space was well preserved, and patella appeared normal

LIHI Treatment (10/9/52 to 11/1/52, 23 days): For treatment of flexion deformity of right knee and hemophilic arthropathies left and right knee

During this admission, flexion contracture of 110 degrees at right knee was corrected by a series of wedge casts until extension to 165 degrees was reached The patient was then fitted for a hemophilia Hessing brace with Lofstrand spring joints at the knee, made to a plaster of Paris model This was accomplished under Avertin anaesthesia Tolserol was also used during this period Increased spring tension was scheduled after patient became ambulatory.

The left leg, which showed only minimal involvement with hemophilic arthropathy, was fitted with a Hewitt-type elastic knee brace with foam rubber padded tongue in one piece Elastic traction was scheduled after patient had become ambulatory for a few days

Examination (12/29/52). Right knee joint: Showed more tendency toward flexion than position of brace joint would allow.

X-ray (12/29/52): Right knee joint: Some irregular calcifications close to medial femoral condyle and in infrapatellar area

Left knee joint Less soft tissue density.

Ambulatory Treatment (12/29/52): Hemophilia Hessing brace was fitted with knee cap with straps and buckles to hold knee down into brace until it was laced Anklet with lacing through two holes in foot plate held ankle down. Tolserol was prescribed to overcome muscle spasm interfering with complete extension.

Examination (1/28/53): *Right knee joint* No soft tissue swelling, effusion, local heat or pain

Left knee joint: Entirely asymptomatic, Hewitt brace discarded

Patient walked with a limp due to fact that right leg had become too long with brace and knee immobilized in almost complete extension

X-ray (4/13/53): *Right knee joint* Some calcification in soft tissues of posterior compartment of joint Considerable improvement since 10/9/52 as knee joint was almost completely extended with a mild degree of posterior subluxation of the tibia Osseous structures showed improved calcification

Ambulatory Treatment (4/13/53 to 4/16/53): Patient received a new hemophilia Hessing brace for right leg and a fiberglass cylinder for immobilization and protection of knee [illegible] ppliances were made to sep.

[illegible] or recurrence of hemorrha[illegible]

Left knee joint: Completely normal

X-ray (10/26/53): *Right knee joint* Held completely extended but there was some posterior subluxation of tibia in relation to femur Articular space of right knee appeared markedly diminished This might be partly due to projection on account of posterior subluxation of tibia Articular surfaces of femoral condyles showed some irregularity and minimal juxta-articular cysts. The patella showed slight signs of the usual deformity but its size did not differ significantly from that of the left.

Impression Patient's condition since April 1953 had been so good and he had been leading such an active life that hemophilic arthropathy of right knee was considered quiescent. Knee should be held in complete extension for at least another six months before attempting to mobilize it.

Ambulatory Treatment (10/27/53): For the right knee, patient received an elastic knee brace extending from above the ankle to below the groin with a foam rubber padded tongue in one piece, continuous lacing from one end to the other, and reinforced on posterior aspect by a heavy spring steel bar to prevent flexion.

Telephone Report (1/27/54): Active flexion considerably improved without loss of extension Elastic brace worn only when patient attended kindergarten Hemophilia Hessing brace was completely discarded

X-ray films (taken out-of-town, 5/19/54; inspected 6/1/54): *Right knee joint* Development of osseous structures almost equaled that of left side No significant enlargement of distal femoral epiphysis Decalcification limited to distal femoral and proximal tibial epiphysis Articular space was well preserved and there was no evidence of further destruction of articular surfaces A small cal-

cium deposit was noted in upper quadriceps pouch. Knee held in almost complete extension with very minimal posterior subluxation of tibia.

Telephone Report **(5/31/54):** Elastic brace worn only at night. Patient was anxious to discard fiberglass brace when swimming. As recent x-ray films showed further rehabilitation of right knee joint, attempt should be made to discard all orthopaedic appliances throughout summer.

Examination **(3/21/56):** *Right calf.* Extensive and painful hemorrhage in January 1956, necessitating prolonged bed rest. There was infiltration but no local heat or pain throughout medial part of musculature of calf.

Right knee joint. AGE 130 degrees.

Right ankle. Moderately swollen with some infiltration of soft tissues on both sides of tendo Achillis. No equinus deformity developed and dorsiflexion remained possible to 90 degrees.

X-ray **(3/22/56):** *Right knee joint.* No evidence of progressive involvement with hemophilic arthropathy.

Right ankle. Moderate changes of hemophilic arthropathy with very slight decalcification of osseous structures. Changes were most marked around os calcis. There was some soft tissue swelling and induration around joint.

Ambulatory Treatment **(3/22/56 to 4/2/56):** A plaster of Paris model was made of right leg to be used in manufacture of a new hemophilia Hessing brace with Lofstrand spring joints at knee and free ankle joint. Thereafter, a wedge cast was applied, holding knee at AGE 120 degrees and ankle at 90 degrees. After three wedgings of cast, knee was held in extension of 170 degrees, and ankle could be brought to 90 degrees. Knee was splinted until orthopaedic appliance was delivered. Patient became ambulatory.

Examination **(4/2/56):** *Right knee joint:* Painless; AGE 170 degrees, AGF 90 degrees.

Right ankle. No swelling but some induration and infiltration of soft tissues around tendo Achillis.

Last Follow-up **(4/3/56):** Improved stance and locomotion.

Case #5737 (Age, 11½) (See Figure 31)

History: Mother's brother died of hemophilia at age 28. Mother's nephew a hemophiliac.

Clinical onset. Early childhood.

Before 1950. *Frequent mild hemorrhages into ankle joints. Bedridden 2 or* 3 weeks for each episode, with complete recovery.

Other joints: Right elbow, asymptomatic with some restriction of extension. *Left knee.* Injured December 1950, hospitalized. Hemorrhage in March 1951; treated with three or four plasma transfusions, unable to walk for a few weeks. Third injury in April 1951, hospitalized for plasma and blood transfusions, thereafter knee became flexed, treated ambulatory; knee became almost straight and patient was able to walk.

September 1951. Injury to left leg, bedridden 10 days, physical therapy was resumed, x-rays taken on 10/29/51 revealed an impacted supracondylar fracture of left femur in healing stage.

Disability at time of first examination: ambulatory with two canes

11/9/51. **Inspection of original x-rays dated 5/29/51.**

Left knee joint Considerable shadow density of soft tissues suggestive of presence of fairly large effusion, probably a hematoma Articular surfaces of femur, tibia and patella appeared normal, but there was some evidence of irregular decalcification throughout distal end of femur as well as proximal epiphysis of tibia

Right knee joint No bone or joint pathology.

11/9/51: **Inspection of x-rays dated 10/29/51**

Left knee joint Marked soft tissue density with well marked capsular thickening, a supracondylar fracture noted in healing stage with periosteal callus formation most marked on lateral aspect. Fracture healing with slight degree of valgus and of flexion deformity An organized hematoma might be present on posterior aspect of distal end of femur Decalcification best seen in medial femoral condyle where it was somewhat suggestive of early cyst formation

First Orthopaedic Examination (11/9/51): *Left knee joint* Moderate enlargement, AGE 140 degrees, AGF 115 degrees; no evidence of abnormal motion at site of supracondylar fracture

Right knee joint Swollen; ballottement of patella indicated fluid, probably blood Local heat and tenderness on pressure throughout joint, AGE 170 degrees, AGF 95 degrees.

X-ray (11/9/51): *Left knee joint* Further healing of supracondylar fracture of femur. Increased shadow density well outlined on posterior aspect of distal end of femur suggestive of an organized hematoma but also of callus formation Soft tissue shadow around knee indicated less swelling and less capsular thickening than on films taken 10/29/51

Right knee joint. Considerable soft tissue density, particularly over medial and posterior aspect of joint Film suggested presence of effusion of moderate degree together with capsular thickening Slight decalcification in one section of medial femoral condyles No evidence of cyst formation or irregularity of articular surfaces.

Ambulatory Treatment (11/13/51 to 1/16/52): For pain and swelling of right knee.

Bed rest and wheelchair ambulation

LHII Treatment (1/24/52 to 1/28/52, 4 days): For flexion deformity of left leg

Examination (1/24/52): *Right knee joint* Minimal swelling

Left knee joint No pain or swelling, AGE 145 degrees, AGF 55 degrees

X-ray (1/24/52): *Left knee joint* The supracondylar fracture of femur had healed with some anterior angulation and slight valgus Fracture line still visualized.

During this hospital admission, a wedge cast was applied from toes to groin, holding knee in maximum extension of 145 degrees In addition, a posterior flexible splint was applied to right knee

Ambulatory Treatment (1/28/52 to 3/13/52): After further wedging, cast for the left leg was removed with knee joint held in extension of 170 degrees. A

plaster model of the left leg was made for a hemophilia Hessing brace with Lofstrand spring joints and a cylinder cast was then applied and worn until the appliance was fitted, with the knee held in complete extension.

After splinting of the right knee, patient was fitted for an elastic appliance similar to a Hewitt-type knee brace but with continued lacing on dorsum of leg and with a solid spring bar in back. This was designed to protect knee from recurrent swelling Patient walked well with new orthopaedic appliances with and without canes

Examination (3/13/52). *Right knee* No swelling; freely movable.

Left knee AGE 170 degrees, AGF 120 degrees Patient to walk as tolerated, without canes if possible

Examination (3/27/52): Patient walked well without canes.

Left knee joint. AGE 170 degrees Lack of complete extension due to mild deformity of distal end of femur resulting from supracondylar fracture Spring tension of Lofstrand joint was increased to maximum.

Right knee joint Slightly swollen.

5/17/52: Patient had returned to school

7/1/52 to 11/4/52: Hemorrhages into elbows, right knee joint and ankles.

Examination (11/4/52): Patient overweight.

Left knee joint No swelling, AGE 180 degrees indicating that deformity from supracondylar fracture had been corrected as femur had grown.

Left ankle. Slight enlargement, no evidence of hematoma, dorsiflexion of foot limited to 100 degrees.

Right knee joint. Almost normal in appearance and function

Right ankle. Slight enlargement due to recent hematoma.

Ambulatory Treatment (11/4/52): Hewitt-type brace for right leg was fitted with regular hinge.

X-ray (11/4/52). *Left knee joint.* Flexion deformity of distal end of femur due to impacted supracondylar fracture was less marked. Juxta-articular cysts were more marked, particularly throughout femoral condyles The tibial epiphyseal line as well as the line of the apophysis at tuberosity of tibia was partly fused in comparison to right side

Right knee joint No bone involvement but some increased soft tissue density due to recent hematoma.

Left ankle and foot: Definite but mild degree of hemophilic arthropathy at ankle with diminished articular space and some roughening of articular surface of tibia and astragalus Skeleton of foot showed some decalcification.

Ambulatory Treatment (12/13/52 to 1/31/53): Patient received a new Hewitt-type brace for right leg

Had been losing weight and walking better

Examination (4/21/53): Patient was again quite overweight. Ambulated very well without cane and without a limp.

Left knee joint· Full range of motion Patient used Lofstrand joints without springs.

X-ray (4/21/53): Left knee joint Slight improvement in recalcification of osseous structures Articular space was well preserved.

Right knee joint: Some widening of diameter of femoral condyles Medial condyle appeared large in relation to lateral condyle

Ambulatory Treatment (5/12/53 to 5/26/53): As left knee joint no longer required a large hemophilia Hessing brace, patient received a Hewitt elastic brace with posterior hinge for the left knee as well as a new Hewitt brace for the right These afforded all necessary support

Examination (7/7/53): Left knee joint Contusion suffered on 6/27/53 was followed by stiffness, swelling and pain Patient received two plasma transfusions and was bedridden for a week. Examination revealed blue discoloration of skin over medial articular space, no swelling or ballottement of patella Tenderness on palpation and pressure was most marked over head of fibula and over medial articular space Extension limited to AGE 170 degrees with definite lateral instability in direction of abduction Patient complained of pain on weight-bearing

Right knee joint Painless but contained some fluid

Ambulatory Treatment (7/7/53 to 7/13/53) To correct recent flexion contracture at left knee and to promote healing of mild injury to the joint

The old hemophilia Hessing brace was reapplied to the left leg with the knee joint locked in complete extension The Hewitt brace was to be worn on the right side as long as there was swelling Patient became ambulatory without a cane and was instructed to discard the hemophilia Hessing brace for the left leg and wear the Hewitt brace if there was no recurrence of pain or flexion contracture.

Examination (8/28/53): Left knee joint Recurrent injuries Patient walked with left leg in external rotation No swelling, AGE 180 degrees, AGF 100 degrees Restriction of internal rotation at left hip was due either to muscular contracture or to deformity of neck of femur.

Ambulatory Treatment (8/28/53): Exercises for left leg

Patient to return to school

Examination (1/12/54): Patient had recently started on cycle of more frequent hemorrhages which required plasma on three occasions He attended school but had to remain at home for several days almost each week

Left elbow Recurrent swelling and hemorrhaging for 5 weeks Moderately enlarged, considerable rigidity AGE 140 degrees, AGF 110 degrees. No local heat Hematoma seemed to be subsiding.

Right knee joint Considerable swelling and ballottement of patella No local heat or pain

Left knee Slight restriction of extension Some irritation of soft tissues in popliteal region due to hinge of Hewitt brace

X-ray (1/12/54): Left elbow. Considerable decalcification of all osseous structures Moderate soft tissue swelling indicated recent hemorrhages Osseous changes limited to slight irregularities of articulation between humerus and ulna

Right elbow. Slight decalcification but no significant bone involvement

Ambulatory Treatment (1/16/54): New Hewitt brace for left leg.

Examination (1/27/54): *Left leg:* Marked swelling, induration and bluish discoloration of calf. Symptoms did not subside after four plasma transfusions for hemorrhage into left calf on 1/14/54. Dorsiflexion at ankle was limited to 150 degrees and knee joint could be extended to 140 degrees. Patient had not borne weight on leg since 1/14/54. During this episode, patient fell and suffered extensive hemorrhages throughout face and a hemorrhage in right orbita was still visible.

LHH Treatment (1/27/54 to 1/31/54, 4 days): For flexion contracture of left knee and the new equinus deformity of left foot.

During hospitalization, a fixed equinus position at the left ankle of 150 degrees was corrected by a wedge cast extending from toes to groin. After two wedgings, dorsiflexion of 115 degrees was obtained.

Ambulatory Treatment (2/4/54 to 3/16/54): A new wedge cast was applied to the left leg from toes to below the knee, holding ankle at 115 degrees. Because of discomfort and swelling on medial aspect of left knee, patient was given a new posterior flexible knee splint and 3 inch Ace bandage to immobilize the left knee joint. The cast was wedged three times, obtained dorsiflexion at left ankle of 90 degrees. Patient was able to bear weight without pain. When cast was removed, the knee joint was no longer swollen and could be extended to 165 degrees. The left calf showed hardly a trace of previous infiltration and induration and was not painful. Dorsiflexion at left ankle was maintained at 95 degrees.

X-ray (3/13/54): *Left foot and ankle:* All osseous structures showed generalized decalcification; some involvement of ankle and proximal tarsal articulations.

A maintenance cast was applied to left leg from toes to below knee holding ankle at 95 degrees. After cast was reinforced and fitted with a Stryker heel, patient was able to return to school and walk as much as tolerated. He also received a new posterior flexible knee splint for the left leg, and a new Hewitt brace for the right knee.

Examination (4/1/54): *Right knee joint:* Hemorrhage into joint on 3/23/54 required several plasma transfusions and bed rest. There was marked enlargement and swelling of joint, with local heat and ballottement of patella. AGE 140 degrees.

Left leg: After cast was removed, leg was painless and could be fully extended. No hardening or pain at calf. Ankle and foot appeared normal with very little restriction of motion. Dorsiflexion to 90 degrees.

Ambulatory Treatment (4/1/54): A posterior flexible splint was applied to the right knee to protect joint and compress effusion. A 3 inch Ace bandage was applied to left leg from toes to below knee. Patient was to remain bedridden until acute condition of right knee subsided.

Examination (4/6/54): *Left leg:* No pain, no swelling of knee or calf, dorsiflexion of ankle possible to 85 degrees.

Right knee joint: Markedly swollen, tenderness on palpation and some local heat, particularly over medial aspect of joint. AGE 140 degrees, AGF 90 degrees. No evidence of continued hemorrhage.

Ambulatory Treatment (4/6/54): Splint was reapplied to right knee. Patient to remain in bed with right knee weighted for greater extension

Examination (4/9/54): *Right knee joint* No pain, AGE 165 degrees Bed rest and weighting continued

Examination (4/15/54): *Right knee joint.* Swollen, slightly warm but not painful AGE 170 degrees.

Left knee joint Asymptomatic

Ambulatory Treatment (4/15/54): Right knee was immobilized in complete extension in the old Hewitt brace which was fitted with a solid steel bar and one-piece tongue Patient was instructed to walk in the house as tolerated

Examination (4/20/54): *Left elbow and forearm* Mild hemorrhage stopped by plasma

Right knee joint: Less swelling, AGE 170 degrees, no pain

Left calf: Considerable atrophy

Left ankle. Dorsiflexion limited to 100 degrees causing patient to walk with left leg in more external rotation than on right side

Patient became cane ambulatory and had returned to school

Examination (5/11/54): Subcutaneous hematoma in right buttock on 5/10/54, moderate hematoma on dorsum of left hand after dog bite.

Left and right knee joints Could be fully extended

Left foot and ankle Condition had improved since exercises for increased dorsiflexion at ankle were started Dorsiflexion possible to 85 degrees

Examination (8/16/54): Patient had long episode of painful hemorrhages into right lower back and abdomen. Remained at home and received eleven plasma transfusions, and Codeine for pain Made a good recovery Walking without canes, wearing Hewitt knee braces

Ambulatory Treatment (8/19/54): A new Hewitt brace was made for the left leg. Patient was instructed in weight-bearing exercises for increased dorsiflexion at left ankle and was to continue resistive exercises to increase strength of musculature of left lower-leg

Examination (12/18/54): For 4 months, patient had been laid up for long periods with stomach hemorrhages requiring 22 plasma transfusions He had also had hemorrhages into right shoulder, right groin and dorsum of right thigh

Right thigh Discoloration of skin and considerable infiltration and induration of subcutaneous fat and possibly of musculature throughout proximal anteromedial half of thigh including groin Hemorrhage into this area had caused development of mild flexion contracture at hip responsible for right-sided limp, poor posture and inability to extend right knee completely

Ambulatory Treatment (12/18/54 to 1/20/55): For flexion contracture at right hip

Patient was instructed to rest 3 hours per day in prone position with 10 lb weight on buttocks Hewitt brace for left leg was to be discarded Patient received new Hewitt brace for right knee with posterior hinge and one-piece tongue.

Examination (4/21/55): Patient had become an increasingly difficult behavior problem He had had gastro-intestinal bleeding and occasional hemor-

rhages into the musculature or the skin and received a number of plasma transfusions. Attended school He was instructed to continue to exercise left foot and ankle to increase dorsiflexion.

Right hip Hardly a trace of flexion contracture.

Examination (5/17/55): Patient's mental condition was becoming more acute. He had been bedridden with new hemorrhages and received transfusions at home.

Left leg Asymptomatic

Right hip No flexion contracture

Right knee joint: Painful in a small area with discoloration of skin from a hematoma and with local heat on dorsum of patella AGE 140 degrees

Right ankle· Dorsiflexion restricted to 100 degrees. The Hewitt brace had been tightly worn and soft tissues distal to the brace were markedly swollen.

Ambulatory Treatment (5/17/55): Patient was fitted with an elastic stocking for right foot and lower-leg The right knee was immobilized on a long, rigid posterior splint to be worn whenever patient was bedridden with hemorrhages and until knee became painless and freely movable.

5/20/55: Patient received transfusion of two units of plasma for goiter-like swelling of neck.

Examination (6/28/55): Gait was much improved Patient to increase activities.

Ambulatory Treatment (7/12/55): Patient was advised to wear elastic stockings on both feet and lower-legs in addition to the Hewitt brace for the right knee.

LHH Treatment (10/18/55 to 11/10/55, 23 days): For acute hemorrhage into popliteal region of left knee and left calf.

During this admission, the hemorrhage was so severe that circulation in left *lower-leg and foot was seriously threatened A series of casts* was required, first for immobilization and protection, and later for correction of flexion contracture of left knee and equinus deformity of left foot. The last cast applied held knee at AGE 175 degrees and ankle in dorsiflexion of 105 degrees

Ambulatory Treatment (11/23/55 *to* 12/8/55): After removal of cast, the left knee joint was normal and in complete extension. Dorsiflexion at left ankle was possible to 95 degrees. Another cast was applied from toes to below knee and a posterior flexible splint was applied with 3 inch Ace bandage to support left knee in complete extension. When cast was removed, left knee was asymptomatic and freely movable. There was no pain or swelling at left ankle and dorsiflexion was possible to 90 degrees with the knee completely extended and to 80 degrees with the knee flexed. There was considerable soft tissue infiltration and induration throughout the medial aspect of left calf but no local heat, redness or tenderness Patient was fitted with a long elastic stocking from toes to mid-thigh Wearing of posterior flexible splint for left knee to depend upon its stability. He was fitted with a spiral-bar lower-leg brace with ankle stop at 100 degrees The old Whitman-type foot plate was used for this brace Patient returned to school, walking with one cane.

Examination (12/22/55): No recurrence of pain or disability at left leg. Need for psychiatric treatment emphasized

Last Follow-Up (4/24/56): Examination for injuries sustained at swimming pool on 3/30/56

Case #6527 (Age, 15)

History: One hemophiliac brother Many bruises, no severe hemorrhages

1950 Right knee joint swollen

Winter, 1953 Right knee suffered first marked hemorrhage Treated with plasma and bed rest Joint did not recover completely Recent recurrence of moderate effusion into the right knee

Other joint Left hip

No orthopaedic treatment

First Orthopaedic Examination (6/18/53): Crutch ambulatory.

Right knee Considerable pathology due to involvement with hemophilic arthropathy Enlargement of joint due partly to small effusion which caused ballottement of patella Medial femoral condyle was quite prominent, tendency toward valgus ACE 170 degrees, flexion was normal

Right thigh and calf. Muscular atrophy

Right leg was conspicuously longer than left, causing pelvic obliquity when patient stood on both legs Right side of pelvis was higher than left and spine formed a postural scoliosis convex toward left side

Pedes Valgi II.

X-ray (6/18/53): *Right knee.* Osseous structures showed most of the characteristic symptoms of hemophilic arthropathy, such as generalized decalcification of all osseous structures, small juxta-articular cysts, enlargement of femoral condyles, particularly medial condyles, plumpness and shortness of patella in comparison with left side Articular space was well preserved but articular surfaces of medial femoral and tibial condyles had suffered moderate damage

Pelvis and hip joints The pelvic obliquity was caused by overgrowth of right leg No significant difference between femoral neck angle on right and left side although angle on right side appeared slightly larger

Ambulatory Treatment (6/19/53 to 6/25/53): Patient received a Hewitt-type elastic knee brace for right knee and a ¾" compensatory lift for the left shoe to correct the pelvic obliquity Walked well

Examination (12/10/53): Patient required 1" compensatory lift on left shoe as he had grown considerably Right knee joint showed slight deformity but was painless and had full range of motion

X-ray (12/10/53): *Right knee* Marked reduction of soft tissue swelling No evidence of fluid in upper quadriceps pouch

Pelvis and hip joints No change in degree of pelvic tilt and the same disparity in femoral neck angle previously noted despite the fact that osseous structures had grown considerably

8/21/54 Severe pain and swelling in left groin, with flexion contracture at left hip Throughout summer, patient was in excellent condition As both legs were covered with mosquito bites, it was possible that a bite on left leg became infected and caused swelling of lymph glands in left groin Crutch ambulatory, unable to bear weight on left leg

Examination (8/26/54): *Left groin* Quite marked swelling of lymph glands and tenderness on palpation

Left hip AGE 140 degrees

There was also some tenderness and possible induration in left lower abdominal quadrant toward wall of pelvis

Diagnosis: Possibility existed of mild hemorrhage into musculature on inside of left ilium although mild infection of glands in the left groin was more probable.

Ambulatory Treatment (8/26/54): Bed rest, application of heat to left groin

Examination (8/31/54): *Left groin.* Less pain, swelling of lymph glands had disappeared Only one small gland was palpable. Left groin was more prominent than right

Left hip Flexion contracture had not improved There continued to be marked tenderness on palpation of left lower abdomen close to iliac crest

Findings were suggestive of previous hemorrhage into musculature on left side of pelvis No indication of continued bleeding

Prescription Tolserol®

LIHI Treatment (9/6/54 to 9/12/54, 6 days): To correct acute flexion contracture at left hip

Traction was applied to left leg which was placed on Braun Boehler lower-leg frame Traction anklet was improvised with foam rubber and flannel bandages Correction of flexion contracture from AGE 140 degrees to 170 degrees was obtained Further treatment to be ambulatory

Examination (9/16/54): Patient walked without limp and flexion contracture of left hip had disappeared To return to school.

Examination (8/25/55): About to enter college Complaints concerned ankles which frequently had tendency toward swelling, pain and occasional stiffness For 3 months, patient had used Ace bandages to stabilize ankles. Reported that there was less pain and disability when feet were held in pronation and dorsiflexion Examination showed both feet in correct static alignment and well formed To stabilize ankles, elastic anklets with closed heels were obtained

Mail Follow-Up (7/20/56). Patient had been well and attending school Hospitalized for removal of two teeth in December 1955.

Case #5926 (Age. 9) (See Figure 61)

History: No family history of hemophilia, brother, age 6, had no hemophilia symptoms

Age 8 Hemorrhage into left knee joint, involvement with hemophilic arthropathy and development of a flexion contracture of almost 90 degrees Several attempts at correction failed

Age 9 Hemorrhage into right knee. Joint was markedly enlarged and painful with mild flexion contracture

Disability at first orthopaedic examination Unable to walk, had not been ambulatory since June 1951, carried by father.

X-ray (3/27/52): *Left knee joint* Marked involvement with hemophilic arthropathy including juxta-articular cysts AGE 125 degrees.

Right knee joint: Hemarthrosis with AGE 120 degrees Films showed generalized decalcification and slight changes in medial femoral condyle and at posterior border of patella suggestive of involvement with hemophilic arthropathy Increased shadow density of soft tissues due to recent hemorrhage.

LIHI Treatment (3/27/52 to 5/4/52, 38 days): For correction of deformities at knee joints. During this hospital admission, a double hip spica cast was applied and wedged on both sides until the left knee was held in extension of 160 degrees and the right knee could be extended to 165 degrees After cast was removed, a plaster of Paris model was taken of left leg for manufacture of a hemophilia Hessing brace with Lofstrand knee joints Thereafter, maintenance casts were applied to both legs from toes to groin and patient started to walk After the large orthopaedic appliance for the left leg was delivered, extension at the knee was increased to 170 degrees At time of patient's discharge from the hospital, right leg, still held in a maintenance cast, was painless and quiescent During this hospitalization, patient suffered a hemorrhage into right forearm for which plasma and ice were administered

Ambulatory Treatment (5/8/52). Right leg was fitted with a Hewitt elastic knee brace.

Examination (6/11/52). *Right knee joint* Swelling, no evidence of hematoma; AGE 140 degrees

Ambulatory Treatment (6/11/52): Cylinder cast applied to right leg

LIHI Treatment (6/26/52 to 6/28/52, 2 days): For correction of recurrent flexion deformity of right knee with AGE 150 degrees

A wedge cast was applied to right leg to obtain 10 degrees increased extension at knee.

Ambulatory Treatment (6/28/52): After wedging of cast obtained AGE 160 degrees at right knee, a plaster of Paris model was made for a hemophilia Hessing brace The right leg was then immobilized in a maintenance cast until delivery of brace.

Examination (8/21/52). Patient had several painful hemorrhages

Right shoulder Pain, slight swelling and local heat but no discoloration of skin.

Ambulatory Treatment (8/21/52 to 9/4/52): Ambulatory plasma transfusions were administered. Patient walked well with new hemophilia Hessing brace

Examination (3/7/53): *Left elbow* Repeated hemorrhages into joint Moderately enlarged, not painful No local heat but some restriction of extension

Ambulatory Treatment (3/7/53 to 3/14/53): Plaster of Paris models were made for new foot sections for the right and left hemophilia Hessing braces, new sandals were delivered

Examination (4/21/53) *Left elbow:* Recurrent pain and bleeding No discoloration of skin but local heat, swelling and tenderness on palpation, flexion and extension

X-ray (4/21/53): *Left elbow* Involvement with mild degree of hemophilic arthropathy Increased soft tissue density at joint and slight involvement with bone pathology at head of radius and at olecranon

Ambulatory Treatment (4/21/53): Left arm was splinted, holding elbow in

extension of 150 degrees. Splint was to be worn only when arm became painful or swollen.

Examination (7/14/53): Condition and function of both knees was excellent with extension possible to 180 degrees. Patient to discard braces when swimming.

Examination (9/8/53): *Left elbow*· New hemorrhage for which a plasma transfusion was required. Joint was moderately swollen with marked local heat and restriction of motion to a few degrees with elbow held at an angle of 140 degrees

Right knee joint. No swelling or pain, active range of motion from ACE 180 degrees to ACF 140 degrees.

Left knee joint: Painless but almost rigid at 170 degrees with more deformity than on right side

Patient continued to walk extremely well with orthopaedic appliances

Examination (2/6/54): *Left knee joint* ACE 175 degrees.

Patient to sleep without braces.

Ambulatory Treatment (2/27/54 to 3/11/54): After plaster of Paris models were made of both legs, new hemophilia Hessing braces were fitted and delivered

Examination (8/14/54): *Right elbow. Recurrent hemorrhages. Little swelling*, some local heat, restriction of extension to 160 degrees with hardly any flexion possible.

Left elbow· Quiescent with some deformity and restriction of motion

X-ray (8/14/54). *Right and left elbows:* Symmetrical involvement of all osseous structures with hemophilic arthropathy

Ambulatory Treatment (8/14/54 to 3/5/55): A dorsal splint was applied to right arm, holding elbow at ACE 155 degrees and forearm in neutral rotation. After swelling had subsided, a plaster of Paris model was made to be used in manufacture of a simple fiberglass brace to support and protect the elbow Patient received brace

Examination (3/5/55): *Right elbow*· Painless, almost rigid in extension of 160 degrees. Pronation, supination and motion at wrist and hand were free. Rigidity was probably due to muscle spasm and disuse.

Examination (9/6/55): *Right elbow.* Very little deformity, no swelling, but complete lack of motion with elbow in extension of 160 degrees Pronation and supination were zero Splint was discarded when it became too small

X-ray (9/6/55): *Right elbow* Osseous changes of hemophilic arthropathy had progressed since 8/14/54 There was still considerable decalcification of all osseous structures While there was no definite evidence of bony ankylosis, there was more cartilage destruction, and very little articular space There was no significant soft tissue swelling Osseous structures had grown since 8/14/54

Examination (9/20/55): *Right elbow* The fact that a small degree of active and passive motion, not exceeding 150 degrees, had recurred spontaneously, improved prognosis for restoration of flexion at elbow

Ambulatory Treatment (9/6/55 to 11/15/55): For increased flexion at right elbow. A plaster of Paris model was made of right arm from metacarpophalangeal

articulations to shoulder, to be used in manufacture of fiberglass appliance to be fitted with an elbow joint with a disk permitting locking of elbow at any degree of flexion. Forces for flexion to be introduced by a bridge in cubital region and by rubber bands

Examination (11/15/55): *Right elbow.* Motion was easier but range of flexion had improved very little

3/27/56: Hemorrhage into right thigh, one unit of plasma administered during hospitalization.

LIHI Treatment (4/17/56 to 4/21/56, 4 days): For recurrence of flexion contracture of right knee due to hemorrhage into right thigh A wedge cast was applied to right leg.

Ambulatory Treatment (4/21/56 to 5/30/56): After a single wedging at the knee, cast on right leg was removed and the hemophilia Hessing brace was reapplied.

Last Follow-up (5/30/56): Patient remained under treatment.

Case #6176 (Age, 5)

History One uncle and one cousin were hemophiliacs.

Clinical onset Infancy

11/1/52: Periarticular hemorrhage at right knee Received 3 units of quick frozen classified plasma.

First Orthopaedic Examination (11/8/52). Patient was bedridden, normal and active child.

Right knee joint Markedly swollen, ballottement of patella indicated fluid Local heat, particularly on dorsum of knee AGE 150 degrees

X-ray (11/8/52): *Right knee joint* Presence of fluid, no evidence of involvement with hemophilic arthropathy

Ambulatory Treatment (11/8/52 to 11/18/52): To prevent development of hemophilic arthropathy

Splinting, unweighting of right knee, and application of ice

Examination (11/18/52). Patient walked and attended school. He was difficult to restrict, and parents felt that knee no longer warranted treatment

Right knee joint: AGE 170 degrees with valgus tendency. No evidence of fluid within joint Splint to be discarded

LIHI Treatment (2/5/53 to 2/11/53, 6 days): For immobilization of right knee and correction of flexion contracture at joint

After recurrent hemorrhages into right knee, patient had been walking with swollen joint held in flexion and had developed a flexion contracture of 150 degrees During this admission, a wedge cast was applied to patient's right leg from toes to below groin and AGE 165 degrees was obtained after first wedging

Ambulatory Treatment (2/11/53 to 5/23/53). *Examination* (2/24/53): *Left knee joint* Considerable swelling and ballottement of patella from recent hemorrhage AGE 130 degrees Further extension caused pain

A second wedging of cast on right leg obtained complete extension of knee joint After removal of cast, a plaster of Paris model was made of leg to be used

in manufacture of a molded leather, jointless Hessing-type brace, to hold leg in almost complete extension and to protect it This appliance was fitted and delivered.

Following a hemorrhage into the left knee, a protective cylinder was applied to the leg to immobilize the knee joint. When cast was removed, AGE 170 degrees had been obtained and the knee was splinted. After the joint became asymptomatic, the patient was permitted to walk as long as the knee remained painless.

X-ray (4/2/53): *Right knee joint·* Soft tissue density was greatly diminished. There was some calcium deposit in suprapatellar region.

Examination (4/9/53): *Left knee joint* Some deformity and enlargement of femoral condyles despite otherwise normal x-ray findings.

Patient received Hewitt-type elastic knee brace with posterior steel spring and complete tongue for left leg. Steel spring bar *in back was reinforced* by a *second spring* for greater support

Examination (9/8/53): Right knee joint. No soft tissue swelling AGE 180 degrees.

Left knee joint. No soft tissue swelling. AGE 180 degrees, AGF 90 degrees

Hewitt brace to be discarded as tolerated and Hessing-type brace for right *leg was to be discarded during night as long as there was no recurrence* of flexion deformity.

Hospital Treatment (11/14/53 to 11/21/53): For hemorrhages around molar which was breaking through. Incisions had to be made. Another tooth was extracted, patient received plasma.

Examination (12/8/53): Patient had discarded Hessing-type brace for right leg as it was too tight at knee causing irritation of skin Walked very well wearing Ace bandage

Right knee joint. AGE 180 degrees, AGF 90 degrees. No local heat or soft tissue swelling. Bony changes, however, required continued use of a brace.

Ambulatory Treatment (12/8/53 to 1/7/54): A plaster of Paris model was made of right leg to be used in manufacture of hemophilia Hessing brace with Lofstrand spring joints at knee. Brace was delivered.

1/7/54 to 2/26/54: Series of hemorrhages into hands and kidneys which required several transfusions

Examination (3/6/54): Patient fell and suffered an extensive hemorrhage into *right knee joint on 2/26/54. Received plasma transfusion because* of pain and marked flexion contracture.

Right knee joint· Moderate swelling with some local heat and tenderness, AGE 140 degrees

Ambulatory Treatment (3/6/54 to 3/13/54): Reapplication of hemophilia Hessing brace; bed rest, ice.

Examination (3/13/54): *Right knee joint:* AGE 180 degrees

Left ankle: Recent hemorrhage. Joint was markedly swollen, particularly *around external malleolus* with discoloration of skin. Motion of left foot was restricted in all directions. There was very little local tenderness.

Ambulatory Treatment (3/13/54 to 4/3/54): A cast was applied to left lower-leg from toes to below knee.

When cast was removed, swelling and tenderness had disappeared and there was no significant restriction of motion A flannel bandage, applied to foot and ankle, was to be worn for several days.

Examination (4/3/54): *Left ankle.* Almost normal with only very slight local heat.

Right knee joint. Greatly improved

Ambulatory Treatment (5/24/54): For large hematoma into soft tissues around right eye after injury Received plasma

9/23/54: Small hemorrhage into left ankle.

LHH Treatment (1/25/55 to 2/5/55, 11 days): For acute hemorrhage into left knee joint.

Examination (1/25/55): *Left knee joint.* Marked enlargement with tenderness on palpation, definite local heat, and some discoloration of skin AGE 120 degrees

A protective cast was applied to left leg from toes to groin, holding knee at AGE 120 degrees Patient received one unit of antihemophilic plasma A second cast was applied holding knee at AGE 140 degrees. During this period, patient suffered from tonsillitis and temperature was elevated There were also hemorrhages into right elbow and forearm, left shoulder and left groin and additional plasma was administered When second cast was removed, knee could be completely extended A splint was applied.

Ambulatory Treatment (2/22/55 to 4/28/55)· A hemophilia Hessing brace was constructed and delivered for the right leg and the right knee joint became fully extended shortly thereafter.

Examination (1/7/56): *Right knee joint* AGE 180 degrees, asymptomatic

Last Follow-up (May 1956): Patient received a new hemophilia Hessing brace for his right leg

Case #6285 (Age, 7)

History: One sister, 14 No hemophilia in family

Age 8 months· Diagnosis established at first signs of subcutaneous and intracutaneous hemorrhages following inoculation for diphtheria and whooping cough

Age 3 Accident, hematoma on left side of neck

Age 4. Fall, injury to left knee, recurrent hemorrhages

Since age 3. Prophylactic treatment with plasma at a hospital, once or twice weekly. No improvement in left knee but little or no bruising or discoloration since beginning of treatment

Schooling Had attended school only 2 weeks

First Orthopaedic Examination (1/31/53): Small, slim child, wheelchair ambulatory.

Left knee joint Enlargement and deformity, definite posterior subluxation of tibia, no soft tissue swelling and no ballottement of patella, AGE 150 degrees, AGF 60 degrees

Left foot. Equinus position of 165 degrees; weight-bearing on metatarsal heads only; most of equinus deformity was due to shortening of tendo Achillis When knee was flexed to 60 degrees, equinus position could be reduced to 120 degrees

Musculature of left leg showed considerable atrophy.

LIII Treatment (2/2/53 to 2/12/53, 10 days): *X-ray* (2/2/53): *Left knee joint:* Increased soft tissue density, some irregularity of articular surfaces and slight thinning of articular cartilage with juxta-articular changes of moderate degree at medial and lateral margins of femoral condyles and at medial tibial condyle. Patella was twice the size of normal right patella Early hemophilic arthropathy

Left foot and ankle. Marked equinus due to soft tissue contracture with very little difference in degree of equinus when knee was flexed or extended. Generalized decalcification but no evidence of hemophilic arthropathy.

During this hospital admission, correction of a marked equinus contracture of left foot and ankle, secondary to hemophilic arthropathy of left knee joint, was started by application of a wedge cast from toes to below knee. A specially constructed hinge with lever and screw attachment was incorporated into this cast and equinus deformity was corrected from 165 degrees to 125 degrees. Cast was extended, during treatment, to include knee in a position of 135 degrees

Ambulatory Treatment (2/14/53 to 8/6/53): After correction of equinus position of left foot as far as cast allowed (110 degrees), cast was wedged at knee for first time while ankle joint was immobilized. After a second wedging at knee, the cast was removed with knee joint extended to 165 degrees Correction, however, of original equinus position of left foot from 165 degrees to 110 degrees was possible only because foot turned into a considerable degree of varus With foot in neutral position between pronation and supination, a considerable degree of correction was lost Since it was more important to hold left foot and ankle in neutral alignment than to force correction of equinus position, a maintenance cast was applied holding knee and ankle in best possible position and foot in neutral alignment. After three wedgings at the ankle, cast was removed with left foot still showing a marked tendency toward equinus and varus position

X-ray (4/16/53): *Left knee joint* No swelling and fairly good calcification, AGE 150 degrees, very little posterior subluxation of tibia in relation to femur.

Left ankle Marked decalcification; equinus deformity had been corrected again to 110 degrees.

Another maintenance cast (to be used for walking) was then applied until a plaster of Paris model was made for a hemophilia Hessing brace without knee joint but with Lofstrand spring joint at ankle to increase dorsiflexion Knee joint was held at AGE 165 degrees. Thereafter, another maintenance cast was applied and worn until hemophilia Hessing brace was delivered, and patient was permitted to walk as much as tolerated Examination on 6/11/53 showed that patient walked well with brace

Examination (8/6/53): *Left ankle* Lofstrand spring joint was highly success-

ful in correcting equinus position of left foot Dorsiflexion was possible to 90 degrees and this position was maintained even without brace

Ambulatory Treatment (8/6/54). A plaster of Paris model was made of left foot and ankle to be used in manufacture of small fiberglass appliance to maintain correction of equinus deformity when patient started swimming

LHH Treatment (8/10/53 to 8/13/53, 3 days): For extensive hemorrhage into posteromedial aspect of right thigh

Examination (8/11/53): *Right thigh:* Considerable infiltration and moderate swelling with beginning discoloration of skin from hematoma into posteromedial aspect.

Right hip Slight degree of flexion and abduction contracture After a transfusion with antihemophilic plasma, bed rest, and application of ice to right thigh, acute symptoms of hemorrhages into thigh as well as minor hemorrhages into back and left arm subsided Patient's general condition was good and temperature was almost normal

The small fiberglass appliance for left foot and ankle, to be worn at night, fitted well, securing foot and ankle in a position of 90 degrees As there was little tendency toward flexion contracture at left knee, patient was permitted to discard hemophilia Hessing brace while in bed

Examination (8/20/53): *Right thigh:* Almost complete recovery

Right hip Slight tendency toward flexion contracture

Examination (9/3/53): *Left knee joint* Mild hemorrhage following contusion There was some puffiness and swelling but no discoloration of skin and no ballottement of patella

Left foot Held at angle of 90 degrees Patient had learned how to exercise foot against resistance of Lofstrand springs.

Examination (9/17/55): Recent hematoma on medial aspect of left upper-arm and to both sides of right elbow Received a transfusion, and hematoma had been subsiding since then There was still slight restriction of extension

Left knee joint Painless, AGE 170 degrees, slight tendency toward recurrence of flexion contracture Patient was sleeping without hemophilia Hessing brace

Left foot Good range of motion with dorsiflexion limited to 90 degrees

Examination (10/24/53) *Left foot* Dorsiflexion limited to 80 degrees

Left knee joint AGE 150 degrees, painless Hemophilia Hessing brace was fitted with knee cap with leather straps to gradually correct flexion contracture

LHH Treatment (11/3/53 to 11/6/53, 3 days): For acute hemorrhage into right thigh causing flexion contracture of hip

After bed rest, refrigeration and three transfusions of antihemophilic plasma, patient made good recovery

Examination (11/14/53). *Right hip* Minimal restriction of extension

Left knee joint AGE 180 degrees Hessing brace lengthened 1 inch and refitted.

Examination (3/13/54): Patient was hospitalized at LHH and given one transfusion for kidney hemorrhage Good recovery

Examination (4/10/54): Left ankle. Freely movable. Dorsiflexion to 85 degrees. Patient was instructed in certain exercises to improve active dorsiflexion and to strengthen weak extensor muscles

LHH Treatment (8/5/54 to 8/7/54, 2 days): For residual contracture of left knee joint and deformity at left ankle.

Examination (8/5/54): *Left knee joint.* No swelling. AGE 165 degrees with a valgus deformity of slightly more than 10 degrees due to accelerated growth of medial femoral condyle. Moderate posterior subluxation of tibia

Left foot: Considerable varus and adductus tendency. Dorsiflexion to 90 degrees with slight degree of convexity of sole.

X-ray (8/5/54): *Left knee joint:* Moderate hemophilic arthropathy.

Left foot and ankle: Some improvement in calcium density. Articular surface of astragalus showed some flattening, probably responsible for persistent restriction of dorsiflexion.

After a plaster of Paris model was made of left leg for a new hemophilia Hessing brace with Lofstrand spring joints at knee and single Lofstrand spring joint for dorsiflexion at ankle, a wedge cast was applied holding knee at AGE 165 degrees and ankle at dorsiflexion of 90 degrees Patient was discharged ambulatory, wearing a cast

Ambulatory Treatment (8/13/54 to 12/18/54): Cast was wedged twice at knee and a maintenance cast was then applied holding knee at AGE 170 degrees and ankle at 90 degrees until hemophilia Hessing brace was delivered Patient walked very well wearing brace. Although foot section maintained dorsiflexion at left ankle to at least 90 degrees, sandal, which was open in back, did not permit simultaneous correction of varus position of heel Treatment was to alternate between further correction of equinus deformity and correction of varus position of heel

Examination (10/7/54): *Left knee joint*· Knee went into flexion when one screw of Lofstrand spring joint of hemophilia Hessing brace became loose. Slight swelling and tenderness. AGE 160 degrees.

A plaster model was made of left foot for a new foot section for the hemophilia Hessing brace with closed sandal to correct lateral deviation of tarsus while ankle was locked at 90 degrees. Although this section was to be worn for a month in place of the previously worn section with Lofstrand spring joint at ankle, it had to be replaced by the old foot section after a short time as heel did not remain in sandal which wrinkled in back and caused irritation. Instead, the heel section was removed and the hemophilia Hessing brace was fitted with an anklet to hold the heel to foot plate This foot plate was worn for four weeks

Examination (12/18/54): Patient walked well

Left foot: Dorsiflexion possible to 80 degrees

Left knee joint AGE 175 degrees

Foot section of brace was exchanged for section with spring joint at ankle for next 4 weeks. During this period of ambulatory treatment, patient had a mild hemorrhage into soft tissues on medial aspect of left thigh He also suffered a hemorrhage into back for which he was admitted to a hospital for transfusions. Good recovery.

Examination (4/2/55): *Left knee joint:* Extension could be increased by removing remaining setscrews from Lofstrand spring joints at knee Complete extension was well tolerated

Examination (9/17/55): *Left knee joint* Asymptomatic, AGE 170 degrees

Left foot Varus deformity of foot, particularly heel, was still quite marked, while equinus component could be corrected to 90 degrees If correction was forced beyond this point, varus deformity increased.

Ambulatory Treatment (9/17/55 to 1/7/56). A wedge cast was applied to left leg from toes to mid-thigh, holding knee in extension of 170 degrees and foot in best possible correction of the equino-varus deformity. Cast was prepared for wedging but not for weight-bearing After cast was wedged twice, it was removed and another cast was applied After a single wedging, cast was removed Left knee could be extended to 175 degrees and exhibited less posterior subluxation of tibia Foot was relaxed and could be corrected into neutral alignment with dorsiflexion of 90 degrees A plaster model of left leg and a separate model of left foot were made to be used for manufacture of a new hemophilia Hessing brace A maintenance cast was then applied until the brace was fitted

X-ray (11/5/55): *Left knee joint* Considerable growth of osseous structures without further destruction of bone or cartilage and with less soft tissue swelling Calcification was somewhat improved

Left foot and ankle: Slightly greater decalcification in mid-tarsal region All bones had grown

After fitting of hemophilia Hessing brace, a maintenance cast was applied from toes to below knee Left knee joint could be completely extended and was supported by a posterior flexible knee splint and 3 inch Ace bandage The new orthopaedic appliance was then delivered and a knee cap added

Last Follow up (3/10/56).

Case #7559 (Age, 10) (See Figure 40)

History: Ancestory negative for hemophilia, brother, age 12, a hemophiliac

Age 8 years Severe polio attack, treated at neighborhood hospital, when treatment was stopped, patient was told to discard orthopaedic appliances Walked with left-sided limp

First Orthopaedic Examination (3/31/55): *Left hip* Musculature was quite weak

Left foot Equino-varus deformity

LIHI Treatment (4/19/55 to 4/30/55, 11 days): For postpolio equino-varus deformity of left foot, causing left-sided limp and requiring correction

During this hospital admission, correction of 20 degrees to dorsiflexion of 110 degrees was obtained by a Quengel cast applied to left leg from mid-thigh to toes A maintenance cast, not intended as a walking cast, was then applied, and patient was discharged wheelchair ambulatory

Ambulatory Treatment (5/7/55 to 6/21/55). After removal of maintenance cast, a wedge cast was applied holding foot at angle of 100 degrees and knee in complete extension Cast was wedged to point where dorsiflexion of left foot was possible to 90 degrees Without weight-bearing, left foot had developed a

tendency toward calcaneovarus rather than previous equino-varus position. A plaster model was made of left foot and lower-leg to be used in manufacture of a spiral-bar lower-leg brace with ankle stop. Leg was then immobilized in a maintenance cast until spiral-bar brace was delivered. Brace was well tolerated and patient became ambulatory.

Examination (7/12/55): *Right elbow.* Recent hemorrhage Moderately enlarged but without local heat or pain This was patient's first joint involvement.

Patient's gait was satisfactory Lower-leg brace to be removed when swimming.

Ambulatory Treatment (7/12/55): *Right elbow:* A protective splint was made to be applied with a 2 inch Ace bandage if and when hemorrhages occurred into elbows.

Examination (9/27/55): Patient was hospitalized for 2 weeks during August and on bed rest at home for another 2 weeks, receiving 14 transfusions for hematuria He also had a mild hemorrhage into right ankle for which he received two transfusions Good recovery.

Right elbow Still slightly enlarged, AGE 160 degrees, no other symptoms

Electromyogram (LIHI 9/28/55): As paralytic foot drop was due to residual weakness of left tibialis anticus and posticus muscles, patient was referred to hospital for electromyogram to determine if muscles could be developed by electric stimulation and exercises Electromyogram revealed definite paralysis of greater part of tibialis anticus Extensor digitorum longus showed partial denervation while all other muscles were normal. Special therapy was not indicated.

Examination (2/3/56): Patient was recently discharged from a two-week hospital admission where he received 13 transfusions for hemorrhages into right knee joint and right thigh Since then, knee again became painful and swollen as soon as he began to walk.

Right knee joint. Markedly swollen, moderately painful Ballottement of patella indicated presence of considerable amount of fluid AGE 170 degrees

Other complaints: Virus infection with gastro-intestinal symptoms and pain in lower left abdominal quadrant.

Ambulatory Treatment (2/3/56 to 4/19/56): Posterior flexible knee splint for right knee.

Examination (4/19/56): Patient was hospitalized frequently between February and April 1956 for internal hemorrhages and hematuria requiring many additional transfusions

Right knee joint Markedly enlarged, AGE 160 degrees.

X-ray (4/19/56): *Knee joints.* Increased soft tissue density indicating synovitis on right side Osseous structures were rather poorly developed and all bones showed considerable decalcification Lower portion of femur and patella appeared larger on right than on left side No evidence of cartilage destruction and little, if any, articular surface irregularity on either side.

Ambulatory Treatment (4/19/56 to 4/26/56): Application of extra long posterior flexible knee splint to right leg, patient to remain wheelchair ambulatory

until condition of right knee had improved and complete extension had been obtained

Examination (4/26/56): Right knee joint Painless but enlarged AGE 170 degrees Splinting was sufficient for immobilization of joint

Examination (5/3/56): Right knee joint Painless, AGE 170 degrees

Splint reapplied, dorsum of right knee to be weighted for long periods of time.

Examination (5/10/56). Right knee joint Completely extended, splint reapplied

Examination (5/19/56): Right knee joint Recurrence of flexion contracture from soft tissue swelling in popliteal region

Ambulatory Treatment (5/19/56). New splint applied to right knee

LHH Treatment (5/24/56 to 6/19/56, 26 days): For hemophilia and acute hemorrhage right lower-leg, hemophilia right knee, and postpolio equino-valgus, left foot

Examination (5/24/56): Patient was pale, restless, in acute pain

Right lower-leg Maximum swelling, skin discolored, taut

Right ankle and foot Slight puffiness but no motor or sensory impairment

After refrigeration and transfusions with whole blood, there was no further pain A protective cast was applied holding right knee in most comfortable position of 90 degrees of flexion and right foot in best possible dorsiflexion of 100 degrees This was repeatedly changed as condition improved Discharged with maintenance cast

Ambulatory Treatment (6/19/56 to 7/5/56): Patient continued to wear maintenance cast on right leg and remained under treatment

Case #6224 (Age, 12) (See Figure 34)

History Mother's father and uncle, patient's brother, age 14, hemophiliacs

Age 1 First hemorrhage when patient cut frenulum of tongue

Age 7 Frequent falls on right knee resulted in hemarthros followed by flexion contracture At first, a short period of traction was sufficient to straighten out the leg Another fall, 3 months later, caused a new flexion contracture, which made walking impossible for one year

Age 9 Patient was treated at a hospital for flexion deformity of right knee A large cast was applied extending from axilla to include right foot After removal of cast, leg was straight A brace, worn for some time, was finally discarded and an attempt was made to restore function to knee by massage and other physical therapy Treatment was successful to the extent that patient could walk without a limp For 2 years, right knee remained asymptomatic and patient was able to attend school and to swim

November 1952 Patient fell on right knee and at same time may have injured right elbow Knee was treated with ice, and swelling and pain gradually disappeared During bed rest, patient developed hemorrhage into right side of neck, followed by muscle spasm Hemorrhage was stopped by transfusion of whole blood

First Orthopaedic Examination (12/8/52): Crutch ambulatory; good general condition.

Right elbow: Moderate enlargement chiefly due to soft tissue swelling Palpation revealed thickening of capsule. Moderate pain on pressure over ulnar aspect of elbow AGE 150 degrees, AGF 70 degrees. Some restriction of pronation and of supination of forearm

Right knee joint· Moderate deformity suggestive of involvement with hemophilic arthropathy. Minimal degree of local heat, very little soft tissue swelling, no ballottement of the patella. Motion limited to AGE 130 degrees, AGF 100 degrees. Definite tendency toward valgus position. Considerable atrophy of musculature of right leg.

LHH Treatment (12/8/52 to 12/22/52, 14 days): For flexion contracture of right knee

X-ray (12/9/52) Right elbow: Mild degree of hemophilic arthropathy with considerable increase in density of soft tissues of joint Slightly generalized decalcification Articular surfaces showed slight irregularities in comparison with left side but no significant thinning of articular cartilage. Juxta-articular cysts were noted in proximal end of ulna and, to a lesser degree, in medial condyle of humerus.

Right knee joint Showed all characteristic signs of a moderate hemophilic arthropathy. Articular surfaces of femoral and tibia condyles were irregular Articular space was diminished but sufficiently well preserved. Femoral condyles showed usual broadening Changes were most marked at patella which was squared off and had lost its distal third There were some cystic areas in patella and very small cysts throughout the proximal epiphysis of tibia. Osseous structures showed decalcification while soft tissue shadow density was increased

During hospitalization, a flexion contracture of the right knee joint with extension limited to 130 degrees was corrected by a wedge cast hip spica until AGE 170 degrees was reached. A plaster of Paris model was then taken of leg to be used for a hemophilia Hessing brace, and leg was immobilized in a maintenance cast applied from toes to below groin. These procedures were performed under Avertin anaesthesia, supplemented with nitrous oxide. Patient was discharged crutch ambulatory, bearing weight on right leg.

Ambulatory Treatment (12/29/52 to 1/9/53): Patient received hemophilia Hessing brace with Lofstrand spring joints at knee for right leg. Crutches were discarded and patient became ambulatory with one cane.

Examination (1/31/53): *Right knee joint*· AGE 170 degrees.

Right lower-leg· Patient instructed in exercises for increasing dorsiflexion and plantar flexion at ankle and for strengthening musculature of lower-leg.

Examination (4/8/53): Patient leading active life, handled orthopaedic appliance very well.

Right knee joint. No recurrence of pain or disability. Still moderately enlarged, and showing original deformity No fluid within joint, no local heat AGE 175 degrees Without brace, patient could flex knee to 140 degrees and stand and walk, but was advised to wear brace most of time.

Right foot Some subtalar rigidity which did not permit supination This was main cause of external rotation of foot

X-ray (4/8/53): Right foot and ankle Moderate generalized decalcification of all osseous structures and some coarseness of trabeculation but no evidence of hemophilic arthropathy.

Ambulatory Treatment (5/27/53 to 6/4/53): Hemophilia Hessing brace for right leg was repaired and measurements were taken for a Hewitt elastic brace with rigid posterior bar and one-piece tongue for right knee for summer use.

Examination (6/18/53): Right knee joint· Hemophilia Hessing brace was discarded, and Hewitt brace worn without recurrence of pain or swelling

Examination (12/10/53): Patient walked without limp Wore Hewitt brace on right leg, but not regularly No difference in length of legs Tendency toward genu valgum on right side and to external rotation of right leg was again noted

Right knee joint Range of motion from AGE 170 degrees to AGF 100 degrees

X-ray (12/10/53): Right knee joint Decrease in soft tissue enlargement and density.

Examination (2/22/54): Internal hemorrhage (2/8/54), for which one whole blood and one plasma transfusion were administered

Fall onto right buttock resulted in large hematoma in medial distal quadrant of right buttock and smaller one over greater trochanter Patient was bedridden When he attempted to walk, he was unable to assume erect posture Ambulated with marked right-sided limp using cane, and had pain throughout right hip. Pale, sick, and had lost considerable weight. Motion at right hip restricted by definite flexion, abduction and external rotation contracture A slight flexion contracture at right knee was secondary to hip

Ambulatory Treatment (2/22/54). Patient was to remain in bed until flexion contracture of right knee and contractures at right hip were corrected Two sandbags were to be placed against lateral aspect of right foot to prevent external rotation To remain prone for at least three 1-hour periods a day with sandbags on right buttock

Examination (2/25/54) Right hip Marked muscular spasticity throughout hip region No evidence of new hemorrhage, and pain apparently did not originate from organized residual hematoma over right greater trochanter and in right buttock

X-ray (2/25/54). Pelvis and hip joints Transverse fracture throughout distal part of femoral neck

LIHI Treatment (2/25/54 to 3/4/54, 7 days)· For fracture of neck of femur without displacement

Under general Avertin anaesthesia, a hip spica was applied to right side, excluding distal third of lower-leg and foot Right hip was held in neutral rotation, 10 degrees of abduction, and complete extension Cast was well tolerated and patient was discharged crutch ambulatory, walking very well

Ambulatory Treatment (3/18/54 to 3/31/54). Cast was reinforced by an old

aluminum spiral-bar hip brace to permit use of an ischial seat strap when walking Cast was removed without recurrence of swelling or pain and knee could be extended to 175 degrees Patient required no further immobilization in cast, activities were to be resumed as tolerated, to attend school after another week

X-ray (3/31/54): *Right hip* Healing of fracture line with good callus formation Fracture line hardly visualized

Examination (4/24/54): *Right leg* Some muscle spasm throughout musculature of right thigh in hamstrings as well as in adductors. No evidence of hematoma Right-sided limp seemed to be caused by pain on posterior aspect of leg close to knee joint Patient to remain off feet as long as pain and muscle spasm continued

X-ray (4/24/54): *Right hip joint* Fracture line in femoral neck was completely obliterated by callus formation, and hardly visible. No evidence of other new pathologic changes.

Examination (1/14/55): *Right wrist* Hemorrhage on volar aspect 2 weeks ago Had moderate pain and could not use hand Most of swelling and infiltration from recent hematoma appeared on volar aspect of forearm just above and at wrist Marked tenderness on palpation and considerable restriction of motion of wrist and fingers Wrist held in flexion of 150 degrees and fingers could not close to a fist. Impairment due primarily to crowding of all structures in tunnel on volar aspect of wrist from hematoma.

Ambulatory Treatment (1/14/55): Patient was given flexible dorsal splint to protect wrist and hand from undesirable motion and permit gradual correction of flexion contracture.

Examination (1/27/55). Poor general condition, pale and depressed.

Patient still complained of pain and disability throughout right wrist and hand Had difficulty writing Examination showed slow but definite progress in healing. Complete recovery without residual contracture expected. Patient was to discard splint

Mail Follow-up (7/20/56). Patient was well and attending school.

Case #6150 (Age, 11)

History: Four generations of hemophiliacs on mother's side.

Cycle Every 3 months, especially during spring and fall

Coagulation time more prolonged in February and early March

At 18 months. Internal hemorrhage into chest.

Right knee and left elbow. Recurrent hemorrhages for 2 years prior to first orthopaedic examination

Patient wore no orthopaedic appliances, had received more than 50 transfusions of whole blood

Attended school until right knee became worse.

First Orthopaedic Examination (10/20/52): Patient was bedridden

Right knee joint Enlarged, due to recent hemorrhage. Ballottement of patella indicated presence of free fluid within joint. ACE 170 degrees, ACF 90 degrees

Right ankle Recent hematoma with considerable restriction of dorsiflexion and of supination.

Left elbow AGE 160 degrees, AGF 70 degrees, pronation and supination were not limited. Patient was left-handed

Pedes plano valgi II/III

X-ray (10/23/52). *Right knee joint.* Enlarged and containing considerable fluid, probably blood, but no evidence of juxta-articular or articular bone pathology.

Left elbow Early changes of hemophilic arthropathy

Ankles Definite involvement with early stage of hemophilic arthropathy. Articular cartilage was thin and there was some irregular destruction of articular surfaces of astragalus, more marked on left side

LIHH Treatment (10/23/52 to 11/8/52, 16 days): During hospitalization, an acute hemarthros of right knee joint, a hematoma at right ankle and rigidity of right foot due to hemophilic arthropathy were treated by bed rest, splinting, refrigeration and plasma The patient received a hemophilia Hessing brace with Lofstrand spring joints at the knee for his right leg and a celluloid plate was prepared for the left foot When discharged, patient was able to stand and walk without discomfort

Ambulatory Treatment (11/13/52) Patient received celluloid plate for left foot

Examination (4/30/53): Right quadriceps was so strong that springs for extension of Lofstrand knee joint were discarded and patient was to sleep without hemophilia Hessing brace

X-ray (4/30/55): *Knee joints* Moderate degree of generalized decalcification of osseous structures Articular surface of medial femoral and tibial condyle of right knee showed slight flattening and irregularity There was evidence of soft tissue swelling and capsular thickening at right knee Soft tissue involvement of right knee was slightly more marked than on films taken on 10/23/52

Ambulatory Treatment (6/30/53 to 7/7/53): Patient received Hewitt-type elastic brace with posterior hinge for right knee to wear throughout summer as tolerated in place of hemophilia Hessing brace The celluloid foot plate for left foot was to be discarded throughout summer

Examination (2/4/54) *Right knee joint* Asymptomatic after periods of recurrent swelling and slight ballottement of patella

Examination (6/3/54): *Right knee joint* Remained asymptomatic after Hewitt knee brace had been discarded for 1 week

Examination (8/26/54). Pain in left hip region Started to walk with limp due to flexion contracture at left hip Some tenderness on palpation of left lower abdominal quadrant Examination suggested mild hemorrhage into iliopsoas muscle preventing complete extension of left hip

Treatment Bed rest and periodic weighting of left buttock Good recovery

Examination (5/31/55): Patient, who had grown considerably, had suffered many internal hemorrhages as well as considerable bleeding into right knee, right ankle and right hip in the 9-month interval between examinations

Right hip: Moderate flexion contracture.

Right knee joint: Restriction of extension to 165 degrees. Some swelling and local heat on dorsum, but no tenderness on pressure. Circumference of right knee was 1 cm. larger than that of left. No pain on weight-bearing.

X-ray (5/31/55): *Right knee joint:* More soft tissue swelling and increased density since 6/30/53.

Ambulatory Treatment (5/31/55 to 6/28/55): Splinting until full extension of right knee joint was restored. Rest for 2 periods of at least 1 hour each day in prone position with 10 lb weight on right buttock to correct flexion contracture at right hip.

Examination (8/31/55): Splint was not worn.

Right knee joint: Considerable pain, marked enlargement with moderate degree of local heat, tenderness over internal and external semilunar cartilage, and slight restriction of complete extension. Increase in degree of external rotation of tibia.

Ambulatory Treatment (8/31/55 to 9/6/55): Splinting and complete rest.

Examination (9/6/55): *Right knee joint:* Less pain and swelling.

After 3 years of home instruction, patient was to return to school for a 6 months' trial period.

Ambulatory Treatment (9/6/55 to 10/13/55): A plaster of Paris model was taken and a new hemophilia Hessing brace for the right leg was made with Lofstrand spring joints at the knee to maintain the knee joint in an asymptomatic condition and enable patient to attend school. Appliance was well tolerated. Knee joint showed definite improvement and patient returned to school. Use of a large orthopaedic appliance was indicated in this case as recurrent attacks of pain *and swelling had occurred after the original hemophilia Hessing brace* had been discarded in July 1953.

The right leg, protected by the new hemophilia Hessing brace, illustrated a marked degree of external rotation of lower-leg, which, in addition to some degree of valgus, originated from unilateral development of a hemophilic knee joint.

Last Follow-up (5/3/56): Attending school, no complaints.

Case #6597 (*Age, 38*)

History: Three brothers who were not hemophiliacs, negative ancestory.

First symptoms when cutting first teeth.

Age 9. *Injury to right knee joint,* followed by recurrent hemorrhages into joint.

Other joints: Elbows, left knee.

Age 28: Appendectomy, survived after many transfusions.

Age 29: Diagnosis of torn cartilage *in right knee joint; arthrotomy performed* which almost cost patient's life. Many transfusions. After operation, right knee joint deteriorated considerably, but in recent years became quiescent and almost painless.

1951-1953: Hospital admissions for hemorrhages into left knee joint.

July 1953: Hospitalized for hemorrhage into right elbow

Orthopaedic appliances. None

Profession. Theatrical agency

Complaints: Recurrent and painful hemorrhages into left knee joint, considerable disability on standing and walking, frequent swelling and pain in right elbow which had increased in recent months and were partly attributable to patient's dependence upon arms for weight-bearing

LIHI Treatment (8/24/53 to 9/11/53, 18 days): *First Orthopaedic Examination* (8/24/53): Good general condition

Left knee joint. Quite painful with local heat, but little swelling and no ballottement of patella AGE 150 degrees, AGF 120 degrees.

Right knee joint: Large, irregular and adherent scar from arthrotomy in 1944 No soft tissue swelling, rigid in extension of 170 degrees with only a few degrees of flexion.

Right elbow: Tenderness and slight soft tissue swelling on radial aspect of joint without significant enlargement AGE 150 degrees, AGF 70 degrees

Left elbow No pain, slight restriction of extension to 160 degrees

X-ray (8/24/53): *Left knee joint:* Soft tissue swelling and increased density, some irregularity, with tendency toward flattening of articular surface of femoral condyles, mild productive changes at margins of patella and at intercondylar notch of femur Suggested synovitis and a mild degree of degenerative joint disease with well preserved articular space Symptoms were not characteristic of hemophilic arthropathy

Right knee joint Quite marked destruction of articular surfaces of patella, femur and tibia with marked thinning of articular cartilage, almost complete disappearance of articular space between patella and femur, flattening of femoral condyle, juxta-articular cysts particularly in center of tibial plateau, generalized decalcification of mild degree Films suggested that greater part of present pathologic changes did not occur during childhood and adolescence but later in life, probably following arthrotomy in 1944

Right elbow: Minimal structural irregularities of all articular surfaces Osseous structures had an almost normal appearance without significant decalcification Suggested an early stage of degenerative joint disease rather than hemophilic arthropathy.

Left elbow. Juxta-articular cystic areas in external condyle of humerus, thinning of articular space between humerus, radius and ulna, spotty decalcification close to articular surface of ulna and in head of radius which appeared slightly larger than on right side These changes may have represented an early stage of involvement with hemophilic arthropathy a long time ago

At outset of this hospital admission, the left leg was treated by means of bed rest, refrigeration, medication and splinting These measures were followed by application of a wedge cast from toes to below groin which corrected the flexion [illegible] ice [illegible] of-

The right knee joint, painless but almost completely rigid, required the protection of a Hewitt-type elastic knee brace.

The right elbow, which had suffered repeated episodes of painful swelling due to small hemorrhages, responded well to transfusions of antihemophilic plasma. Patient received a fiberglass jointless brace for protection of this joint.

Examination (12/7/53): *Right elbow* Recurrent pain on posterolateral aspect and some loss of motion. At least 3 days each week, elbow became swollen, painful and stiff Fiberglass brace did not afford much help although it was worn two-thirds of time Palpation revealed some thickening of capsule and tenderness on pressure but no local heat AGE 150 degrees, AGF 90 degrees Very little restriction of pronation and supination

Knee joints Asymptomatic, left knee was in complete extension

X-ray (12/7/53): *Right elbow* Slight degree of spotty decalcification throughout lateral condyle of humerus, particularly close to articular surface

Diagnosis: Neither x-ray nor clinical findings were suggestive of hemophilic arthropathy at right elbow This was possibly synovitis and an early stage of atrophic arthritis independent of hemophilia. Recurrent painful swelling may have been related to use of cane. Treatment. Butazolidin®, and Aspirin if necessary

Examination (4/5/54): *Left knee joint.* Asymptomatic. Occasional steps taken without brace had not produced tendency toward flexion contracture, patient to gradually discard hemophilia Hessing brace and to be provided with a Hewitt elastic knee brace with well-padded, one-piece tongue and solid steel bar in back

Right elbow. Painless, range of motion from AGE 160 degrees to AGF 80 degrees, musculature of arm in excellent condition, fiberglass splint had been discarded

Right knee joint Hewitt elastic knee brace had been discarded

Patient was well standardized with Butazolidin®.

Ambulatory Treatment (4/5/54 to 4/6/54): Fitting and delivery of Hewitt elastic knee brace for left knee

Examination (8/16/54): Patient no longer required hemophilia Hessing brace for left leg. The Hewitt elastic knee brace without hinge was well tolerated Lower extremities remained asymptomatic and patient walked quite fast occasionally using one cane.

Right elbow Occasional pain and swelling without evidence of hemorrhage. Minimal soft tissue swelling existed at posterolateral aspect. Flexion was restricted to 90 degrees

LIHI Treatment (8/17/54 to 8/24/54, 7 days): For extraction of tooth Avertin anaesthesia, no hemorrhaging, good recovery

Examination (1/10/55): Continued with Butazolidin® although patient felt that medication was hardly required

Knees Asymptomatic, patient walked well without orthopaedic appliances No hemorrhages.

Left knee AGE 170 degrees, AGF 150 degrees

Right knee AGE 170 degrees, AGF 155 degrees

Right elbow Occasional pain, no swelling, no significant deformity AGE 140 degrees, AGF 90 degrees Musculature of arm in excellent condition Patient was advised to wear fiberglass brace when pain occurred

Last Follow-up (June 1956): Letter reported that patient was walking very well without orthopaedic appliances and had remained free of hemorrhages and pain Routine orthopaedic examination was scheduled for September 18, 1956.

Case #6003 (Age, 17) (See Figure 56)

History: Family history negative for hemophilia

Circumcision Bleeding for several weeks

Age 5 Laparotomy

Age 7 Hemorrhages into left knee and left ankle

Other joints Left elbow, shoulder and wrist, right knee and ankle

Internal Abdominal, right ear

Cycle Spring and fall

Had all common childhood diseases except measles

Treatment At least one hospital admission per year, total of 100 transfusions

Orthopaedic treatment Open lengthening of right Achilles tendon, traction for right leg, drop foot night splints These did not prevent development of deformities Since age 10, patient wore Klenzak brace with drop foot spring on left leg.

Schooling: Tutored at home

Disability: Wheelchair ambulatory, completely ambulatory for only 1 week during 2 years prior to first orthopaedic examination

First Orthopaedic Examination (6/7/52): *Right shoulder and elbow* Aside from recent hemorrhages into subcutaneous tissues, function was normal and musculature was well developed

Right wrist Swelling and pain, joint was thin with restriction of dorsiflexion and volar flexion as well as minimal limitation of supination Due to recent hemorrhage into right wrist, patient did not use his hand and resisted passive dorsiflexion.

Left shoulder Involvement with hemophilic arthropathy and recurrent subluxation Elevation restricted to 160 degrees, abduction to 90 degrees

Left elbow. Moderate deformity due to hemophilic arthropathy with extension limited to 125 degrees Flexion was normal Supination of left forearm restricted to 50 degrees Flexors were much stronger than extensors

Left wrist Painless but showed no active dorsiflexion Grip of hand was weak Fifth finger was stiff

Right knee joint Marked involvement with hemophilic arthropathy, AGE 130 degrees, AGF 70 degrees

Left knee joint Almost freely movable and painless, AGE 175 degrees, AGF 70 degrees

Left foot Marked equino-varus deformity with almost complete ankylosis. Musculature of left lower-leg showed advanced atrophy

Right foot: Equino-varus deformity similar to that on left side despite open lengthening of tendo Achillis Slight motion at right ankle. Atrophy of musculature of lower-leg not as marked as on left side.

LHH Treatment (6/24/52 to 8/29/52, 66 days): For flexion contracture at right knee joint, pain at right wrist, multiple hemorrhages

X-ray (6/30/52): *Right wrist and hand·* Carpal bones and distal articular surface of radius showed minimal but definite irregularities, probably due to minute hemorrhages into bones. Little generalized decalcification

During this hospitalization, a wedge cast was applied to right leg from toes to below groin, holding knee in extension of 145 degrees. Foot was placed in neutral position with as much dorsiflexion as marked and rigid equinus deformity permitted After five wedgings of cast, a plaster model was made of leg for a hemophilia Hessing brace with Lofstrand spring joints at the knee and without ankle joints Thereafter, a maintenance cast was applied holding knee at AGE 160 degrees A plaster model was also made of the left leg to be used in the manufacture of a hemophilia Hessing brace with Lofstrand spring knee joints. Knee remained fully extended but no correction of the equino-varus position of left foot was possible Subsequent examination indicated that left ankle and foot were more relaxed than when original model was taken. Foot was held in good longitudinal alignment with long axis of leg and a new model of foot and ankle was taken Flexion contractures at knee joints were fully corrected and patient was discharged wearing two hemophilia Hessing braces with built up foot sections completely accommodating the marked equino-varus deformity on both sides Patient was able to stand and walk with two canes and with no pain on weight-bearing.

Patient also received Sterling Bunnell's active splints for dorsiflexion of wrists. Wrists, hands and fingers, with exception of left fifth finger, became freely movable and painless.

Elbows became painless, and flexion contracture which limited extension at left elbow to 150 degrees was of little significance to left arm. Patient's left elbow was fitted for an active correcting wire splint built like a Sterling Bunnell wrist splint

Patient was advised to correct slight flexion contracture at right hip (150 degrees) by resting daily in prone position

General condition was greatly improved.

Examination (1/24/53): Patient walked well without canes or crutches, although he had some difficulty balancing body when standing.

Right knee joint AGE 170 degrees, no range of motion. Quadriceps was still so weak that brace had to be worn with springs at knee in maximum tension. No attempt to flex the joint to be made.

Left knee joint AGE 180 degrees, AGF 90 degrees. Quadriceps greatly improved

Right elbow· Freely movable.

Left elbow· Range of motion from AGE 130 degrees to AGF 60 degrees. Function within this range was painless.

2/20/53. Hemorrhage into left knee joint was not treated with plasma immediately, bedridden 3 months, complete recovery

7/13/53: Hemorrhage into right lumbar region, plasma administered immediately; quick recovery

Examination (8/4/53): Driving car. Extensive use of brace for left elbow resulted in greater range of motion. Left wrist and hand were asymptomatic and left thumb was no longer numb Patient was able to stand on right and left leg alone without support. Preferred small crutch for balance when walking outdoors. General condition was good

Right arm: Normal function

Left shoulder Elevation and abduction showed defect of 30 degrees

Left elbow· Range of motion from AGE 140 degrees to AGF 45 degrees, an increase of 25 degrees

Left forearm· Pronation normal; supination showed defect of one-third of normal range.

Left wrist and hand Wrist almost free, fist was powerful with exception of fifth finger which did not participate.

Right knee joint AGE 180 degrees, no flexion Right quadriceps showed improvement

Left knee joint Range of motion from AGE 170 degrees to AGF 120 degrees, recent hemorrhage had decreased range of motion

Hips No pain and no significant restriction of motion

X-ray (8/4/53). Pelvis and hip joints Symmetrical and normal development of osseous structures of pelvis Right acetabulum somewhat flatter than left Femur showed lack of development, more marked on left side Head of right femur showed slight deformity. No evidence of hemophilic arthropathy

Right knee joint· Moderate generalized decalcification of all osseous structures. Shaft of fibula unusually thin Patella was plump, showing shortening and broadening of its distal pole Articular space was greatly diminished with some destruction of articular cartilage and some juxta-articular cyst formation of slight degree in external femoral condyle Distal end of femur showed slight valgus deformity due to overgrowth of medial femoral condyle

Left knee joint Same degree of generalized decalcification as on right side Articular space was much better preserved indicating that joint would remain freely movable provided hemorrhages could be prevented Fibula was thin but less so than the right. Patella showed normal size but some defect in articular surface opposite femoral condyles. There was very little structural change suggestive of hemophilic arthropathy in femoral and tibial condyles, but tiny cysts were well visualized

Right foot and ankle Marked generalized decalcification of all osseous structures, almost complete ankylosis of ankle joint and markedly diminished articular spaces throughout tarsal and metatarsal structures Foot showed equino-varus deformity of highest degree (almost 180 degrees) and ankle joint showed some valgus position due to overgrowth of region of medial malleolus

Left foot and ankle Same degree of equinus position of 180 degrees as on

right side Osseous structures were better calcified, particularly above ankle, and ankle joint was better preserved than on right side Less marked varus deformity of tarsus and metatarsus than on right foot.

Comment: Clinical and x-ray examinations showed steady progress in rehabilitation of stance and locomotion which could not have been originally anticipated

Examination (8/9/54): *Small hemorrhages into right knee joint and right* elbow in past year

Orthopaedic appliances had been so successful that canes were completely discarded. Patient drove car, worked regularly as auto salesman while continuing as honor student at college As he could not stand or walk without orthopaedic appliances because of equino-varus deformity, he should eventually have small appliances corresponding in construction to lower-leg and foot sections of hemophilia Hessing braces he was wearing. These provided such satisfactory protection and stability that patient wished to continue wearing them although his control over his knee joints indicated that they were no longer necessary.

August 1954 to December 1955: Hemorrhages into right elbow; left palm, left ankle, *also gastro-intestinal bleeding*

Examination (12/19/55): *Knees* Asymptomatic with good stability and complete extension at right knee, slight restriction of complete extension but flexion to almost 90 degrees at left knee *Function was so good that hemophilia Hessing* braces could be discarded. Musculature of both thighs was improved.

Ankles and feet No change in deformity.

Ambulatory Treatment **(12/19/55 to April 1956):** Plaster models were taken for lower-leg Hessing-type braces without ankle joint and with same platform support for equino-varus deformity of both feet. Braces, delivered by mail, were not correctly fitted.

Last Follow-up (6/4/56): Patient returned for correct fitting of lower-leg Hessing-type braces

Case #6531 (Age, 11)

History: *Only child, no known case of hemophilia in family.*

Age 4 months Hospitalized for hemorrhages in right arm. Other joints involved Elbows Internal hemorrhages Kidneys, nose.

Age 8 years Patient fell on left knee, hospitalized for 11 weeks and treated with traction and casts, discharged wearing two double-bar caliper braces with ring locks at knee

First Orthopaedic Examination (6/19/53): Patient was leading active life; school, rough play on streets Crutch ambulatory wearing double-bar caliper leg braces Good general condition Stood and walked just as well without braces which did not fit

Right elbow Slight restriction of extension and flexion

Right forearm Minimal restriction of supination

Musculature well-developed, no pain.

Left knee joint. Slightly enlarged with flexion contracture of 145 degrees with

posterior subluxation of tibia and moderate valgus deformity Could be flexed to 55 degrees.

Right knee joint: Range of motion from 155 degrees to 55 degrees

Left lower-leg Considerable degree of external rotation probably originating from knee joint

Right foot Marked valgus position Internal malleolus was quite prominent and covered with a small bursa due to pressure from brace

X-ray (6/23/53): Knee joints Thinning of all articular cartilages and considerable irregularity of articular margins There was very little juxta-articular cyst formation Slight soft tissue swelling noted about both knees Early stage of hemophilic arthropathy, slightly more extensive on right side.

Right elbow Slight involvement with hemophilic arthropathy

Left ankle. Some involvement with hemophilic arthropathy, with marked valgus position of astragalus

Right ankle Osseous structures appeared almost normal with exception of valgus position

***LHH Treatment* (7/7/53 to 7/21/53, 14 days):** Patient was treated for flexion contractures and hemophilic arthropathies of both knee joints After plaster models were made of legs to be used for manufacture of hemophilia Hessing braces, wedge casts were applied on both sides Extension of right knee was increased from AGE 160 degrees to AGE 175 degrees, that of left knee from AGE 150 degrees to AGE 170 degrees Right knee was then immobilized on a posterior knee splint After braces were fitted and delivered, patient was discharged ambulatory without crutches or canes

***Examination* (8/11/53):** Patient walked well

***Examination* (3/15/54):** Patient reported mild nosebleed, contusion of right elbow, and severe contusion of right knee joint, followed by hemorrhaging

Right knee joint Moderate swelling, no ballottement of patella, slight degree of local heat, no significant pain, AGE 140 degrees

***Examination* (4/19/54):** *Right shoulder:* Following acute hemorrhage into joint, there was marked swelling, discoloration of skin, local heat

Right knee joint Still moderately swollen, no evidence of fluid in joint, heat or pain AGE 150 degrees

Treatment for right shoulder Plasma, bed rest, icebags To remain at home

***Examination* (5/7/54).** Patient was still unable to attend school

Right knee joint: Superficial bruises over medial femoral condyle AGE 150 degrees

***Ambulatory Treatment* (5/7/54)** Lofstrand spring joint of right hemophilia Hessing brace was locked at AGE 150 degrees, and fitted with knee cap to hold knee securely in position and enable patient to bear weight on leg

***Examination* (5/25/54):** *Right shoulder* Marked rigidity Motion hardly possible to more than 40 degrees of elevation and of abduction Local heat throughout joint.

Right knee joint As patient felt that knee could be extended well beyond AGE 150 degrees, lock screw was removed

Left forearm and wrist Slight discoloration of skin and puffiness throughout wrist and hand Pronation and supination markedly restricted and quite painful.

***LHH Treatment* (5/25/54 to 5/29/54, 4 days):** For new hemorrhages into right shoulder and left forearm and wrist.

X-ray (5/26/54): *Right shoulder* Slight decalcification of head of humerus but no involvement with hemophilic arthropathy.

Left forearm and wrist. No abnormal findings.

Knee joints Improved calcium density of all osseous structures and no soft tissue swelling.

Ankles: Considerable valgus position, more marked on left side, with decreased calcium density of all osseous structures. No evidence of articular involvement or bone destruction.

As patient's clotting time was fairly rapid, he received a transfusion with antihemophilic quick frozen plasma Right shoulder and left wrist became less painful. After instruction in exercises for both arms to improve range of motion at right shoulder and restore free supination and pronation to left forearm, he was discharged wheelchair ambulatory

Examination (8/16/54): *Right knee joint:* Suffered contusion. Joint, which had been completely extended, showed AGE 150 degrees with moderate swelling and patient was wheelchair ambulatory. Knee cap to be worn with hemophilia Hessing brace to restore complete extension

Examination (8/26/54): *Right knee joint* AGE 165 degrees; less swelling, patient had started to walk.

Examination (9/7/54): *Right knee joint* AGE 150 degrees. Patient received attachment for elastics to increase extension.

Examination (9/21/54): *Right knee joint* Extension possible to 165 degrees. Patient to attend school.

Examination (5/7/55): Patient walked well

X-ray (5/12/55): *Knee joints:* Better calcification, no further destruction

Examination (8/25/55): *Left ankle:* Recent hemorrhage.

Left knee joint AGE 165 degrees, spring joint locked in maximum extension [illegible] to be used

[illegible] no longer required

[illegible] eived a new hemophilia Hessing brace for right leg.

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CHART I

Case No.	Age at First Examination*	Clinical Onset of Hemophilia				Family History of Hemophilia			Cyclical Hemorrhages		Internal Hemorrhages			Previous Orthopaedic Treatment		First Major Joint Involvement						Total Major at First Examination	
		Birth to 1 yr	1-5 yr	5-12 yr	Other	Yes	No	Other	Yes	No	Yes	No	Only After First Exam	Yes	No	Knee	Elbow	Ankle	Hip	Shoulder	Multiple	Knees	Elbows
5682	15	x					x			x	x			x							x	2	2
5736	19			x			x			x	x				x	x						1	1
6531	11	x					x			x	x			x			x					2	2
5672	43				x		x			x		x			x						x	2	2
5624	2	x					x			x		x			x	x						2	
7559	10	x				x				x			x	x			x						1
7513	9	x					x			x		x		x		x						1	
5830	6	x					x		x		x			x		x						1	1
5737	11	x				x				x			x		x			x				1	1
3597	4	x					x			x	x			x		x						1	
5733	34		x			x				x			x	x							x	2	2
7185	7	x					x			x		x			x						x	1	1
5653	6	x					x			x		x		x		x						1	
6527	15	x				x				x		x			x	x						1	
5926	9			x			x			x		x			x	x						2	
6224	12	x				x				x	x			x		x						1	1
6294	25	x				x				x	x				x				x			1	
7207	7	x						x		x	x			x							x	2	2
6269	24		x			x				x		x		x		x						2	2
7032	51	x				x				x	x			x		x						2	2
6678	9	x					x			x	x			x		x						2	2
6597	38	x					x			x		x			x	x						2	2
7146	8	x				x				x	x				x			x					1
7389	8	x					x			x	x			x							x	2	2
6809	7	x					x			x		x			x	x						2	
7762	18	x					x			x		x		x			x					2	1
7769	23	x				x			x		x			x							x	2	2
5732	33		x			x			x		x			x							x	2	2
5396	23	x				x				x		x			x					x		1	
6949	10	x					x			x	x			x		x						1	
6504	10	x				x				x			x		x	x						2	1
6140	6	x					x			x			x	x							x	2	
7301	7	x				x				x	x				x						x	2	
6150	11		x			x			x		x				x	x						1	1
5112	14	x					x		x		x			x		x						2	2
6004	14		x			x				x	x				x	x						2	2
6264	4	x					x		x			x			x			x					
6271	16	x				x				x	x				x						x	2	
6607	6	x				x				x	x			x		x						2	1
6028	26		x			x			x		x			x							x	1	1
6295	7	x					x			x	x				x	x						1	
6131	4	x					x			x		x			x	x						2	
6311	47			x			x			x			x		x	x						1	1
6287	15	x					x			x			x		x						x	2	2
6495	35	x				x				x	x				x		x					2	2
5747	13	x				x				x	x			x		x						2	2
6003	17	x					x		x		x			x		x						2	1
5637	11	x				x			x		x			x		x						1	2
6516	22		x				x			x	x				x						x	2	2
5566	14	x				x				x	x				x	x						2	
6175	5	x				x				x			x		x	x						1	
5316	24		x			x				x	x			x		x						2	2
8058	26	x				x				x	x				x	x						2	2
6975	45	x				x				x			x		x						x	2	2
8182	17	x					x			x	x			x		x						2	
7994	5	x					x			x			x	x							x	2	2
Totals		44	8	3	1	27	28	1	9	47	32	14	10	28	28	31	4	3	1	1	16	88	60

*First Orthopaedic Examination

Joint Involvement					Joints Involved After First Examination							Total Joint Involvement							Degree of Disability at First Examination				Locomotion Disability at First Examination				
Ankles	Shoulders	Wrists	Hips	Total	Ankles	Elbows	Knees	Hips	Shoulders	Wrists	Total	Knees	Elbows	Ankles	Shoulders	Hips	Wrists	Total	Total	Marked	Moderate	None	Bedridden	Wheel Chair	Crutch	Cane	Difficult
2	2	2	2	12							–	2	2	2	2	2	2	12	x				x				
1				3			1				1	2	1	1				4	x				x				
1				5					1	1	2	2	2	1	1		1	7			x				x		
1				6							–	2	2	1				6		x				x			
				2							–	2						2	x				x				
				1			1				1	1	1					2			x			x			
		1		2	1						1	1		1			1	3	x				x				
				2	1	1		1		1	4	1	2	1		1	1	6		x			x				
2				4		1	1		1		3	2	2	2	1			7		x						x	
				1	2		1				3	2		2				4	x				x				
2		2		8								2	2	2			2	8		x					x		
				2	1		1				2	2	1	1				4	x				x				
				4	2	2					4	1	2	2				8	x				x				
			1	2								1				1		2			x				x		
				2		2			1		4	2	2		1			5	x				x				
				2				1		1	2	1	1			1	1	4		x					x		
		1	1	3	1						1	1		1		1	1	4		x					x		
2				6								2	2	2				6		x					x		
2				6								2	2	2				6			x				x		
2				6							–	2	2	2				6	x				x				
	1			5							–	2	2		1			5	x				x				
				4							–	2	2					4		x					x		
2				3							–		1	2				3	x				x				
				4	2						2	2	2	2				6		x					x		
1				3							–	2		1				3	x				x				
				3							–	2	1					3	x					x			
	2		1	7								3	2		2	1		7		x						x	
				4								2	2					4		x					x		
	1			2				1			1	1			1	1		3		x					x		
				1	1						1	1		1				2			x						x
1				4								2	1	1				4	x				x				
2				4								2		2				4		x					x		
1				3				1			1	2		1		1		4		x						x	
1				3	1			2			3	1	1	2		2		6	x				x				
				4				1			1	2	2			1		5	x				x				
				4				1			1	2	2			1		5		x					x		
1				1	1	2	1				4	1	2	2				5			x					x	
1				3					1		1	2		1	1			4	x				x				
1				4						1	1	2	1	1			1	5		x					x		
				2			1	1			2	2	2					4			x					x	
				1	1	1		1			3	1	1	1		1		4		x				x			
				2	1						1	2		1				3	x				x				
			1	3			1				1	2	1			1		4			x						x
1				5						1	1	2	2	1			1	6	x				x				
				4								2	2					4		x					x		
2	1		1	8					1		1	2	2	2	2	1		9		x				x			
2	1	1		7			1				1	2	2	2	1		1	8	x					x			
				3								1	2					3		x					x		
2		1		7							–	2	2	2			1	7				x				x	
				2							–	2						2	x					x			
				1	1	1	1		1		4	2	1	1	1			5	x				x				
	2			6							–	2	2		2			6		x					x		
				4								2	2					4		x					x		
2				6						2	2	2	2	2			2	8		x						x	
1				3							–	2		1				3		x				x			
				4							–	2	2					4		x			x				
36	10	8	7	209	16	12	9	9	6	7	59	97	72	82	16	16	15	295	22	25	8	1	21	8	19	7	2

LHH* Admissions						Hospital Treatment										
Total Admissions	No. Days	Quengel Cast	Wedge Cast	Protective or Maintenance Cast	Total Casts	Plaster of Paris Models	HHB‡	Fiberglass Brace	Foot Plate	Hewitt Elastic Knee Brace	Splint	Double- or Spiral-Bar Brace	Total Appliances	Quengel Cast	Wedge Cast	Protective or Maintenance Cast
2	31	3	9		12	2(L),1(U)	1	1(L)					1(L) 1(U)			
2	95					1	1						1			
2	18		2		2	2	2				1		3			
—	—															
1	25		1	1	2	1						1	1			
2	37	1		2	3										1	1
1	13	2(U)†			2(U)	1										1(U)
1	31	1(U)	1		1(U) 1(L)	1	1						1			1(U)
3	31		3	1	4						1		1		1	3
1	1															
2	50		1		1	4	2		2		1		5			
3	41			2	2	6(L) 1(U)	5	1(U)	1				8(L) 1(U)			
4	33			6	6											3(L) 2(U)
1	6															
3	44		4	3	7	1	1						1			1
2	21		1	2	3	1										
—	—															
2	43	1		2	3	3(L) 2(U)	4	2(U)			2		6(L) 2(U)			
—	—															
1	32			1	1	2(L),1(U)	2	1(U)			1		3(L),1(U)			
2	10	2	1	1	4	2									1	1
2	25		1		1	1(L) 1(U)	1	1(U)		2	2		5(L) 1(U)			
—	—															
3	16	1	3		4											
—	—															
1	3			2	2	1	1				3		4			
—	—															
1	11		1		1	2	2						2			
2	47			3	3	1	1						1			
—	—															
3	38	1			1	1	1						1			
1	12	1		1	2											3
1	1			2	2											
1	16					1	1				1		2			
4	98		1	1	2	1	2						2			
1	39	1	1	1	3	2	1				2		4			
1	10			2(L) 1(U)	1(L) 1(U)											3
2	55		1	1	2	1	1				1(L) 1(U)		2(L) 1(U)			
4	34	1	1	1	3										1	3
—	—															
2	19		2		2	1		1					1		3	5
1	23		1		1	1	1			1			2		1	1
—	—															
4	160	5	6	2	13	10	8	2			2(L) 2(U)		12(L) 2(U)			
3	50	1	1		2	2	2				2		4			
1	8	1			1									2	2	1
1	66		1	1	2	2	2				4(L)		2(L) 4(U)			
5	100	2	1	1	4						1		1	1	1	1
—	—															
1	21			1	1	1	1			1			2			
2	17		1	2	3						1		1			2
—	—															
1	29	1	1		2	2	1			1			2			
—	—															
1	23		1	1	2	3	2						2			6
1	3			1	1											
Totals		21(L)† 3(U)†	47	43(L) 1(U)	111(L) 4(U)	64(L) 6(U)	48	3(L) 6(U)	3	5	21(L) 7(U)	1	81(L) 13(U)	3	13	34(L) 4(U)
88	1508	24	47	44	115	70	48	9	3	5	28	1	94	3	13	38

*Lenox Hill Hospital

†(U) refers to upper extremities, (L) refers to lower extremities. Where no letter appears, the cast or appliance relates to the lower extremities.

‡HHB refers to hemophilia Hessing brace.

Additional Ambulatory Treatment: Total Casts	Plaster of Paris Models	HHB	Fiberglass Brace	Foot Plate	Hewitt Elastic Knee Brace	Splint	Double- or Spiral-Bar Brace	Total Appliances	Ambulatory Treatment Only: Quengel Cast	Wedge Cast	Protective or Maintenance Cast	Total Casts	Plaster of Paris Models	HHB	Fiberglass Brace	Foot Plate	Hewitt Elastic Knee Brace	Splint	Double- or Spiral-Bar Brace	Total Appliances
	2	3						3												
	1	1						1												
	1	1						1												
																		2		2
2	1					3(L) 1(R)	1	4(L) 1(R)												
1(R)	1(R)	1	1(R)					1(L) 1(R)												
3(R)	3(L) 4(R)	3	4(R)			4(R)		3(L) 8(R)												
4	2	1			8	2	1	12												
									1	1	6	8	16	10	1	4	4	5	2	25
3(L),2(R)	4	2		2	1		2	7												
					1			1												
1	4(L) 2(R)	3	2(R)		1	2(R)		4(L) 4(R)												
		1			1	1(R)		2(L) 1(R)												
													2	1			1	2	1	5
													2	2						2
2	3(L) 1(R)	3	1(L) 1(R)		1	1		6(L) 1(R)												
											1	1	5	4		1				5
	4	4						4												
											1	1	3(L) 1(R)		1(R)	2			1	3(L) 1(R)
													1	1			1			2
													2	1		1	1			3
	1	1			2	2		5												
3	5	2		3				5												
	1	1			1			2												
	2	1		1	1	2		5												
	4	8				1		7												
	4	2		2	2			6												
3	3			2		3	1	6												
	1	1						1												
4	2(L) 2(R)	2	2(R)			1(R)		2(L) 3(R)												
													1(R)	1(R)			2			2(L) 1(R)
8	3	3						3												
2	3	2	1		1	1		5												
													1(L) 1(R)	1	1		1			3
3	5(L) 2(R)	3	1(L) 2(R)	1		3(R)		5(L) 5(R)												
	2	2						2												
2	4	4						4												
													2	1		1				2
						1		1												
2	4	3			1	2		7												
													2	2						2
													1	1						1
6	1	1				1		2												
49(L) 4(R)	70(L) 12(R)	59	3(L) 12(R)	11	21	19(L) 12(R)	5	117(L) 24(R)	1	1	8	10	37(L) 3(R)	21(L) 1(R)	2(L) 1(R)	9	10	9	4	58(L) 2(R)
53	82	59	15	11	21	31	5	141	1	1	8	10	40	23	3	9	10	9	4	60

	Over-all Treatment												
	Quengel Cast	Wedge Cast	Protective or Maintenance Cast	Total Casts	Plaster of Paris Models	HHB	Fiberglass Brace	Foot Plate	Hewitt Elastic Knee Brace	Splint	Double- or Spiral-Bar Brace	Total Appliances	Rehabilitation at End of First Active Phase of Treatment
	2	9		11	4(L) 1(U)	4	1(U)					4(L),1(U)	Wheelchair amb., 1 HHB, 1 wedge
					2	2						2	Amb., 1 HHB
		2		2	3	3				1		4	Amb., 2 HHB's
										2		2	Relief of pain at night, 2 splints
		1	1	2	1						1	1	Amb., 1 double-bar brace
	1	1	3	5	1					3(L),1(U)	1	4(L),1(U)	No change
	2(U)		1(U)	3(U)	1(L) 1(U)	1	1(U)					1(L),1(U)	Cane amb., 1 HHB
	1(U)	1	1(U)	1(L),2(U)	4(L),4(U)	4	4(U)			4(U)		4(L),8(U)	Amb.; 1 HHB
		4	4	8	2	1			8	3	1	13	Amb., 1 HHB, 1 elastic knee brace
	1	1	6	8	16	10	1	4	4	5	2	26	Amb., 1 HHB
		1		1	4	2		2		1		5	Amb., 1 HHB
			2	2	6(L) 1(U)	5	1(U)	1				6(L),1(U)	Amb.; 1 HHB, 1 fiberglass foot plate
			9(L),2(U)	9(L),2(U)	4	2		2	1		2	7	Amb.; 1 HHB
									1			1	Amb., 1 elastic knee brace
		4	4	8	5(L) 2(U)	4	2(U)		1	2(U)		5(L) 4(U)	Amb., 1 HHB, 1 elastic knee brace
		1	2	3	1	1			1	1(U)		2(L),1(U)	Cane amb., 1 HHB
					2	1			1	2	1	5	Amb., 1 HHB
	1		2	3	4(L) 2(U)	4	2(U)			2		6(L),2(U)	Amb., 2 HHB's
					2	2						2	Amb., 2 HHB's
			1	1	2(L) 1(U)	2	1(U)			1		3(L) 1(U)	Cane amb., 2 HHB's
	2	2	2	6	5(L) 1(U)	3	1(L),1(U)		1	1		6(L),1(U)	Crutch amb., 2 HHB's
		1		1	1(L) 1(U)	1	1(U)		2	2		5(L),1(U)	Cane amb., 1 HHB, 1 e k b*
			1	1	5	4		1				5	Amb., 1 HHB
	1	3		4	4	4						4	Amb., 1 HHB
			1	1	3(L) 1(U)		1(U)	2			1	3(L),1(U)	Amb., 1 spiral-bar brace
			2	2	1	1				3		4	Crutch amb., 1 HHB
					1	1			1			2	Amb., 1 HHB, 1 elastic knee brace
		1		1	2	2						2	Amb., 2 HHB's
			3	3	1	1						1	Amb., 1 HHB
					2	1		1	1			3	Amb., 1 HHB
	1			1	2	2			2	2		6	Amb.; 1 HHB
	1		4	5	5	2		3				5	Cane amb., 1 HHB
			2	2	1	1			1			2	Amb., 1 elastic knee brace
					3	2		1	1	3		7	Amb.; 1 HHB, 1 foot plate
		1	1	2	8	8				1		9	Amb. between parallel bars, 2 HHB's
	1	1	1	3	6	4		2	2	2		10	Amb. with walker, 2 HHB's
			4(L) 1(U)	4(L) 1(U)	3			2		3	1	6	Amb., 1 foot plate
		1	1	2	2	2				1(L),1(U)		3(L) 1(U)	Crutch amb., 1 HHB
	1	2	4	7	2(L) 2(U)	2	2(U)			1(U)		2(L) 3(U)	Amb., 1 HHB
					1(U)	1(U)			2			2(L),1(U)	Amb.; 1 elastic knee brace
		5	5	10	4	3	1					4	Amb., 1 HHB
		2	1	3	4	3	1		2	1		7	Amb., 1 HHB, 1 elastic knee brace
					1(L) 1(U)	1	1		1			3	Amb., 1 HHB
	5	6	2	13	10	8	2			2(L) 2(U)		12(L) 2(U)	Amb. between parallel bars, 2 HHB's
	1	1		2	2	2				2		4	Crutch amb., 2 HHB's
	3	2	1	6	5(L) 2(U)	3	1(L) 2(U)	1		3(U)		5(L) 5(U)	Crutch amb., 2 HHB's
		1	1	2	4	4				4(U)		4(L),4(U)	Cane amb., 2 HHB's
	3	2	2	7	4	4				1		5	Cane amb., 1 HHB
					2	1		1				2	Amb., 1 HHB
			1	1	1	1			1	1		3	Amb., 1 HHB, 1 elastic knee brace
		1	4	5	4	4			1	3		8	Amb., 1 HHB
					2	2						2	Amb., 1 Hessing type brace, 1 HHB
	1	1		2	2	1			1			2	Amb.; 1 HHB 1 elastic knee brace
					1	1						1	Amb., 1 Hessing-type brace
		1	1	2	3	2						2	Amb., 2 HHB's
			7	7	1	1				1		2	Amb.; 1 HHB
Totals	25(L) 3(U)	59	85(L) 5(U)	169(L) 8(U)	171(L) 21(U)	130(L) 1(U)	8(L) 19(U)	23	36	49(L) 19(U)	10	256(L) 39(U)	
	25	59	90	177	192	131	27	23	36	68	10	295	

*e k.b. refers to elastic knee brace.

CHART I—Continued

Time Span	*General Orthopaedic Rehabilitation at Date of Last Contact*	*Over-all Care*	*Reduction in Size, Type or Number of Major Orthopaedic Appliances*
21 days	Completely amb., 2 HHB's, school, job, drives car	4½ yr	
1 mo.	Complete rehabilitation	2¾ yr	
11 days	School	3 yr	
10 days	Declined orthopaedic treatment	10 days	
25 days	Amb., school	5 yr	
1½ mo	Under treatment	15 mo	
18 days	Cane amb	10 mo	
1 mo.	School, swims	4½ yr	
2 mo.	School	4½ yr	Amb., 1 elastic knee brace, 1 spiral bar brace
3½ mo.	Amb., 2 HHB's, school, swims	10 yr	
22 days	Amb., 2 HHB's, job, drives car	4½ yr	
21 days	Amb., 2 HHB's, school	1¾ yr	
26 days	School, swims	5 yr	
8 days	School	2 yr	
1½ mo	Amb., 2 HHB's, school, swims, under treatment	4 yr	
1 mo.	School, active life	4 yr	
1 mo.	Amb., 1 elastic knee brace and spiral bar brace, job, car	3¼ yr	HHB discarded for elastic knee brace except for unusual activities
3 mo	School	1¾ yr	
25 days	Job	6 mo.	
3 mo	Completely amb., job, travels (death from cerebral hemorrhage)	20 mo	
1½ mo	Completely amb., swims	2½ yr	Amb., 1 HHB, 1 elastic knee brace
18 days	Completely amb. without appliances, active life	3 yr	Appliances discarded
12 days	Swims	16 mo	
12 days	School, under treatment	15 mo	
2 mo	School	2½ yr	
1 mo	Cane amb., 1 HHB, desk job, swims without HHB	9 mo	
12 days	Complete rehabilitation, (death from abdominal hemorrhage)	7 mo.	
11 days	Job, drives car	3½ yr	2 HHB's worn only for heavy work, long trips
30 days	Amb	8 mo	
3½ mo	School, swims	1½ yr	
12 days	Amb., 1 HHB, 1 elastic knee brace	2¾ yr	
1 mo	Completely amb., school	3½ yr	HHB discarded
8 days	Amb., 1 HHB, 1 elastic knee brace, school, under treatment	1½ yr	
16 days	School	3½ yr	
2½ mo.	Cane amb., college, drives car, swims	6 yr	
1 mo.	Amb. without appliances, college	4 yr	Appliances discarded
4 days	Under treatment	3½ yr	
1½ mo	Amb	3½ yr	
3½ mo.	School	2 yr	
1 day	Amb. without appliances, job	1 yr	Appliances discarded
4 mo	Swims	3 yr	
23 days	Swims, active life	3½ yr	
1 mo.	Amb., 1 HHB, 1 elastic knee brace, job	3½ yr	HHB discarded on alternate days
3 mo.	Amb., swims	3½ yr	
1 mo.	Amb., job, drives car	2 yr	
4 mo	Amb., school	4½ yr	Amb. with 1 HHB, 1 fiberglass knee brace
2 mo.	Amb., college, job, drives car	4 yr	
5 mo.	Amb., school	5 yr	
13 days	Able to continue job	2 yr	HHB discarded
21 days	School	25 days	
3 mo.	School	3½ yr	
3½ mo	Complete rehabilitation, (death from cerebral hemorrhage)	1 yr	
1 mo.	Job, active life	2½ mo.	
1 mo.	Complete rehabilitation, unable to work for other reasons	1½ yr	
23 days	Under treatment	3 mo.	
1½ mo.	School	6 mo.	

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